

**STUDY OF MANAGEMENT OF ADVANCED CASES
OF
JUVENILE NASOPHARYNGEAL ANGIOFIBROMA**

Dissertation submitted for

M.S Degree examination

BRANCH – IV- OTORHINOLARYNGOLOGY

Upgraded Institute of Otorhinolaryngology

Madras Medical College

Chennai-03



**The Tamilnadu Dr.M.G.R. Medical University
Chennai**

ACKNOWLEDGEMENT

I would like to thank Dr.T.P.Kalaniti M.D., Dean, Madras Medical College,Chennai for permitting me to pursue my study in the institution .

I wish to place my sincere thanks and gratitude to the Director of Upgraded Institute of Otorhinolaryngology,Madras Medical College,
Prof. S. Ammamuthu M.S.,DLO., for his kind guidance and encouragement during the period of this study without which the study would not have found its present form.

My sincere thanks to Professor U.Venkatesan M.S.,DLO., for his valuable suggestions and guidance in this study.

I wish to place my immense gratitude to Prof . A.K. Sukumaran M.S.,DLO., additional professor of upgraded institute of otorhinolaryngology for his constant encouragement and guidance throughout the period of this study.

I also extend my sincere thanks to Prof.S.Kulasekaran M.S., DLO.,additional professor of upgraded institute of otorhinolaryngology for his valuable suggestion and support during this study.

I also would like to extend my warm gratitude to all our Assistant professors who have been helpful in all possible ways in making this dissertation see the light of the day.

Finally I would like to extend my heartfelt gratitude to all the patients who have taught us all we know..

CONTENTS

Introduction	- 1
Aim	- 3
Review of literature	- 4
Materials and methods	- 30
Inclusion and exclusion criteria	- 31
Proforma	- 33
Observation	- 58
Discussion	- 60
Conclusion	- 67
Bibiliography	- 71

INTRODUCTION

Juvenile nasopharyngeal angiofibroma (JNA) is a benign, locally aggressive, highly vascular tumour often occurring at the superior margin of the sphenopalatine foramen occurring exclusively in adolescent males.

JNA is uncommon and accounts for approximately 0.5% of all head and neck neoplasms.

The median age at diagnosis is 15 years. Presenting symptoms commonly include nasal obstruction and epistaxis. Approximately 20% of patients have evidence of skull base invasion at the time of diagnosis. JNAs may be hormonally dependant. Hagen et al studied androgen receptor binding in cultured tumour fibroblasts from 3 patients with JNA and demonstrated that cell proliferation increased with testosterone and decreased with antiandrogens.

Spontaneous regression is unlikely but has been observed with increasing age . The main stay of treatment of JNA is surgical excision. Radiotherapy is useful in achieving local control in inoperable cases. There are limited data

suggesting that chemotherapy may be efficacious for the occasional patient with an incompletely resected tumour in whom RT has failed to achieve local control.

This study has been conducted in the Upgraded institute of Otorhinolaryngology, Madras medical college, Chennai-03 during the period 2005 to 2008. The following is a discussion of the management of patients with JNA and, in particular, the assessment and management of advanced cases of JNA.

AIMS OF THE STUDY

- 1.To study the age distribution,clinical presentations of JNA.
2. To Study and apply various Radiological investigations for assessment of the exact extent and vascular supply of locally advanced cases of JNA.
3. To choose appropriate management modality based on the extent of the tumour.
- 4.To assess the effectiveness and complications of various treatment modalities and to explore ways to minimize complications and optimize the outcome of treatment.

REVIEW OF LITERATURE

The history of JNA dates back to Hippocrates, who described removal of a 'hard nasal polyp' using a midline nasal splitting incision. Harma presented a comprehensive of the subject in 1958 and noted that piecemeal removal of the tumour through the external nares was done till 1834. Chelius(1834) advanced the understanding by reporting that less bleeding was encountered when the tumour was removed along with its roots(Vascular pedicle).In 1906,Chaveau coined the term juvenile nasopharyngeal angiofibroma.

THEORIES OF ORIGIN OF JNA

Ringertz(1938)

The tumour arose from the periosteum or the nasopharyngeal vault.

Som and Neffson(1940)

Inequalities in the growth of the bones forming the skull base resulted in hypertrophy of underlying periosteum in response to hormonal influence.

Bensch and Ewing (1941)

The tumour probably arose from the embryonic fibrocartilage between basi-occiput and basi-sphenoid.

Brunner (1942)

Originated from Pharyngobasilar and Buccopharyngeal fascia.

Osborn (1959)

These were either hamartomas or residues of fetile erectile tissues which were subject to hormonal influences.

Girgis and Fahmy (1973)

Postulated the tumour to be a paraganglioma. Existence of paragangliomatous tissues around the terminal part of internal maxillary artery in the pterygopalatine fossa of stillborn infants.

Angiofibromas may arise from vestiges of atrophied stapedia artery, although it is not possible to validate such assertion.

Hughes

Angiofibroma arises from remnants of craniopharyngeal duct.

Wills

Angiofibroma is the result of inflammatory immune response.

Stenberg

Angiofibroma is a type of hemangioma.

Martin (1948)

Use of testosterone caused atrophy of the tumour.

Schiff (1959)

Combination of oestrogen and testosterone caused atrophy of tumour.

Muller (1999)

Transforming growth Factor Beta-1 may play a role in the stromal cell proliferation and angiogenesis associated with JNA.

Hwang and Patterson

Presence of Androgen receptors in Angiofibromas explains the unique clinicopathologic features of these tumours.

Reham-Schimd (1998)

Expression of CD-34 antigen in nasopharyngeal angiofibroma indicates the increased proliferative potential of endothelium of small vessel component of angiofibroma.

Lloyd G Howard 1999

Recurrence of angiofibroma was due to the residual mass in the pterygopalatine fossa and invasion of sphenoid bone.

This can be differentiated radiologically by persistence of widening of distance between root of pterygoid and posterior wall of maxilla in cases of recurrences from pterygopalatine fossa.

Recurrence from the mass invading sphenoid shows pushing of root of pterygoid anteriorly towards maxilla and reducing the distance between root of pterygoid and posterior wall of maxilla.

Susan C. Abraham, Elizabeth A. Montgomery,

Francis M. Giardiello, and Tsung-Teh Wu (2001)

Frequent occurrences of JNA in cases of Familial Adenomatous Polyposis(FAP) due to frequent beta-catenin mutations in JNA.

SURGICAL ANATOMY

Site of origin

- Described by Neal.
- Periosteum of the posterosuperior aspect of sphenopalatine foramen.
- At the junction between sphenoidal process of the palatine bone and the pterygoid process of the sphenoid bone.

Mode of spread

- Spreads submucosally along preformed anatomical pathways through foramina and fissures.

Anterior

- Spreads to the nasal septum, lateral nasal wall.
- Involves the ethmoid sinus, erodes the posterior wall of the maxillary sinus and involves it.

Posterior

- Involves the anterior wall of sphenoid sinus and spreads into the sinus.
- Erodes the pterygoid plates.

Lateral

- Pterygopalatine fossa and may present in the cheek.
- Extends further laterally into the infratemporal fossa.

Inferior

- Nasopharynx and postnasal space.

Superior

- To orbit via the inferior orbital fissure and then superior orbital fissure and middle cranial fossa.

Intracranial extension

- seen in 10 % of the cases
- mostly extradural
- The tumor reaches the cranial vault through three paths.

Two lateral pathways(lateral to carotid and cavernous sinus)

1. Through superior orbital fissure
2. Directly through the greater wing of sphenoid from pterygopalatine fossa and Infratemporal fossa.

Medial pathway(medial to the carotid and cavernous sinus)

-through the sphenoid sinus and sella turcica making surgical excision difficult.

-Intradural involvement is rare,when present is a contraindication for surgery.

PTERYGOPALATINE FOSSA

Boundaries:

Anterior- Superior aspect of posterior surface of maxilla.

Posterior- Root of pterygoid process and adjoining part of

Anterior surface of the greater wing of sphenoid.

Medial - Upper part of perpendicular plate of palatine bone.

Orbital and sphenoidal process of the palatine bone also take part.

Lateral - Opens into infratemporal fossa, through the pterygo-Maxillary fissure.

Superior- Undersurface of body of sphenoid.

Inferior - Closed by the pyramidal process of palatine in the angle between the maxilla and the pterygoid process.

Communications:

Anterior-With the orbit through the medial end of the inferior orbital fissure.

Posterior- Middle cranial fossa through the foramen rotundum.

- Foramen lacerum through the pterygoid canal.

- Pharynx ,through the palatovaginal canal.

Lateral- Infratemporal fossa through the pterygomaxillary fissure.

Medially- With the nose through the sphenopalatine foramen.

Inferiorly- With the oral cavity through the greater and lesser Palatine canals.

Contents:

- Third part of maxillary artery and its branches.

- Maxillary nerve and its two branches zygomatic and posterior alveolar.

- Pterygopalatine ganglion and its numerous branches

Containing fibres of the maxillary nerve mixed with autonomic fibres.

SPHENOPALATINE FORAMEN

Boundaries:

Superiorly- Body of the sphenoid

Anteriorly- By orbital process of palatine bone.

Posteriorly-Sphenoidal process of palatine bone.

Inferiorly -Perpendicular plate of palatine bone.

Oval in shape. Located at the posterior end of the superior meatus(85%) or the middle meatus(5%) or is bridged by the basilar lamina of the middle turbinate (10%).

Sphenopalatine vessels emerges out from pterygopalatine fossa, through the foramen into the nasal cavity.

INFRATEMPORAL FOSSA

Roof:

Medially by the Infratemporal surface of the greater wing of the sphenoid and by a small part of the temporal bone.

Laterally:

Roof is incomplete where the infratemporal fossa communicates with the temporal fossa through the gap deep to the zygomatic arch.

Roof is pierced by the foramen ovale and by the foramen spinosum.

Floor: is open.

Medial wall:

Formed by the lateral pterygoid plate and the pyramidal process of the palatine bone.

Lateral wall:

Formed by the ramus of the mandible.

Anterior wall:

Formed by the infratemporal surface of the maxilla and by the medial surface of the zygomatic bone.

Anterior and medial walls are separated in the upper parts by the pterygomaxillary fissure through which the infratemporal fossa communicates with the pterygopalatine fossa.

Upper end of the pterygomaxillary fissure is continuous with the anterior part of the inferior orbital fissure through which the infratemporal fossa communicates with the orbit.

Posterior wall: is open

Contents of the infratemporal fossa:

1. Muscles- Medial and lateral pterygoid,temporalis.
2. Vessels - Maxillary artery and its five branches.
3. Nerves - Mandibular division of fifth cranial nerve.

TEMPORAL FOSSA

Boundaries:

- Above - Temporal line of frontal bone.
- Below - Upper border of zygomatic arch laterally and by infratemporal crest of greater wing of sphenoid bone medially.
- Anterior wall- Zygomatic bone and parts of sphenoid and frontal bones.
- Floor - 'H' shaped suture where four bones (frontal, Parietal,sphenoid,temporal) join.

MIDDLE CRANIAL FOSSA :

It is shaped like a butterfly, narrow and shallow in the middle and deep on each side.

Boundaries:

Anterior :

- Posterior border of lesser wing of the sphenoid.
- Anterior clinoid process and anterior margin of Chiasmaticus.

Posterior :

- Superior border of petrous temporal bone.

Floor :Dorsum sellae of sphenoid.

Lateral :

- Greater wing of sphenoid.
- Antero-inferior angle of parietal bone.
- Squamous temporal bone.

Median – Body of sphenoid

Lateral - Cerebral surface of greater wing of sphenoid

- Anterior surface of petrous temporal bone
- Cerebral surface of squamous temporal bone

CONTENTS:

Median area :

Sulcus chiasmatis - leads to optic canal.

- Optic canal
- Bounded laterally by lesser wing of sphenoid.
 - In front and behind by two roots of lesser wing.
 - Medially by body of the sphenoid.

- Sella turcica
- Contains tuberculum sellae in front
 - Hypophyseal fossa in the middle
 - Dorsum sellae behind

Lateral area :

Lodges the temporal lobe of the brain.

SUPERIOR ORBITAL FISSURE:

Boundaries

- above by the lesser wing of sphenoid
- below by the greater wing of sphenoid
- medially by body of sphenoid

Foramina in greater wing of the sphenoid:

Foramen rotundum:

- leads anteriorly to the pterygopalatine fossa.
- lies posteroinferior to medial end of superior orbital fissure.
- Transmits maxillary nerve.

Foramen ovale :

- lies posterolateral to foramen rotundum and leads inferior to the infratemporal fossa.
- Transmits mandibular nerve, lesser petrosal nerve, accessory Meningeal artery, emissary vein connecting the cavernous sinus with pterygoid plexus of veins.

Foramen spinosum:

- Lies posterolateral to foramen ovale
- Leads inferiorly to infratemporal fossa
- Transmits middle meningeal artery, meningeal branch of mandibular nerve, posterior trunk of middle meningeal vein.

Emissary sphenoidal foramen(foramen of Vesalius):

- Transmits emissary vein connecting cavernous sinus with the pterygoid plexus of veins.

Groove for the middle meningeal vessels:

-Leads forwards from foramen spinosum.

Foramen Lacerum:

-Lies posteromedial to foramen ovale, lower part of canal filled with cartilage.

-No significant structure passes through it except for meningeal branch of the ascending pharyngeal artery and an emissary vein from the cavernous sinus.

-Upper part of the canal is traversed by the internal carotid artery.

- in upper part of the foramen, the greater petrosal nerve unites with the deep petrosal nerve to form the nerve of pterygoid canal, which leaves the canal by entering the pterygoid canal in the anterior wall of foramen lacerum.

ORBIT

Inferior orbital fissure:

Transmits,

1. Maxillary nerve
2. Zygomatic nerve
3. Orbital branches of pterygopalatine ganglion
4. Infraorbital vessels
5. Communication between inferior ophthalmic veins and the pterygoid plexus of veins.

Superior orbital fissure :

Lateral part

- lacrimal nerve
- frontal nerve
- trochlear nerve
- superior ophthalmic vein
- meningeal branch of lacrimal artery

Middle part

- Upper and lower divisions of the oculomotor nerve
- Nasociliary nerve
- Oculomotor nerve
- Abducent nerve

Medial part

- Inferior ophthalmic vein
- Sympathetic nerves from plexus around the internal carotid artery.

CAVERNOUS SINUS

- Large venous sinus located in the middle cranial fossa,on either side of the body of sphenoid bone.Its interior is divided into number of spaces by trabeculae.
- Floor of sinus is formed by endosteal dura .
- Lateral wall,roof and medial wall is formed by meningeal dura
- Anteriorly it extends upto medial end of the superior orbital fissure.
- Posteriorly upto apex of petrous temporal bone.

Structures in the lateral wall of cavernous sinus:

Occulomotor nerve

Trochlear nerve

Ophthalmic nerve

Maxillary nerve

Trigeminal ganglion

Structures passing through the centre of the sinus:

Internal carotid artery

Abducent nerve

Structures in the sinus are separated from blood by

Endothelial lining.

Incoming channels:

From orbit:

- Superior ophthalmic vein.
- Branch of inferior ophthalmic vein.
- Central vein of retina.

From brain:

- Superficial middle cerebral vein.
- Inferior cerebral veins from temporal lobe.

From meninges:

- Sphenoparietal sinus
- Frontal trunk of meningeal vein.

Draining channels :

- Transverse sinus through the superior petrosal sinus.
- Internal jugular vein through the inferior petrosal sinus and through a plexus around internal carotid artery.

- Pterygoid plexus through emissary veins passing through foramen ovale, foramen lacerum, emissary sphenoidal foramen.
- Facial vein through superior ophthalmic vein
- Right and left cavernous sinus communicate with each other via the anterior and posterior intercavernous sinuses and through the basilar plexus of veins.
- All these communications are valveless communications and blood can flow through them in either direction.

INTERNAL MAXILLARY ARTERY

Larger terminal branch of external carotid artery given off behind the neck of mandible .

Divided into three parts (by the lateral pterygoid muscle)

First part(Mandibular part) - runs along the lower border of lateral pterygoid muscle.

Second part(Pterygoid part) - runs either superficial or deep to the lower head of lateral pterygoid muscle.

Third part(Pterygopalatine part) – passes between two heads of the lateral pterygoid and through the pterygomaxillary fissure to enter the pterygopalatine fossa and lies in front of the pterygopalatine ganglion.

Branches:

First part:

1. Deep auricular
2. Anterior tympanic
3. Middle meningeal

4. Accessory meningeal

5. Inferior alveolar

Second part:

1. Deep temporal

2. Pterygoid

3. Masseter

4. Buccal

Third part:

1. Posterior superior alveolar

2. Infraorbital

3. Greater palatine

4. Pharyngeal branch

5. Artery of pterygoid canal

6. Sphenopalatine artery

EXTERNAL CAROTID ARTERY

- Branch of common carotid artery
- Lies anterior to internal carotid artery
- Begins in the carotid triangle at the upper border of thyroid cartilage.

- Terminates behind the neck of mandible into maxillary and superficial temporal artery.

Branches

Anterior

Superior thyroid, lingual, Facial.

Posterior

Occipital, posterior auricular.

Medial

Ascending pharyngeal .

Lateral

Maxillary, Superficial temporal.

PATHOLOGY OF JNA

Gross appearance

Firm, rubbery lobulated swellings, nodularity increases with age. Nasopharyngeal portion which is covered by mucosa appears pink and extrapharyngeal portion appears white or grey.

On section

Tumour has reticulated, whorly or spongy appearance and lacks a true capsule. Edge however is sharply demarcated and easily distinguishable from surrounding structures (Pseudo capsule).

Microscopy

- composed of vascular and stromal components
- Vascular component predominates in early lesions
- Stromal component predominates in long standing cases.
- Vascular pattern is composed of large thin walled sinusoidal vessels lined by flattened epithelium, unsupported by a muscular coat and the closer the vessels to the surface the smaller they become.
- In long standing cases, there is tendency towards gradual compression of sinusoids so that the lining endothelial cells

are pushed against each other like cords ,while in others intravascular thrombosis occurs.

- The stroma is composed of interlacing bundles of collagen in which stromal cells are seemed to radiate outwards from the vessels and in which localized areas of myxomatous degeneration may be observed.

Immunohistochemistry

Androgen receptor has been detected in the stromal cells of JNA. Antiandrogen hormonal therapy has been used preoperatively to suppress JNA growth and enhance the resectability of the tumor and minimize bleeding during surgery.

Familial adenomatous polyposis (FAP) results from germline mutations in the adenomatous polyposis coli (APC) gene that subsequently alters the [beta]-catenin signaling pathway. In addition to a high tendency for the development of colorectal neoplasms, patients with FAP develop JNA 25 times more frequently than an age-matched population. Recently, activating [beta]-catenin mutation without the APC gene mutation has been reported in sporadic JNA.

Expression of various growth factors, such as transforming growth factor [beta]1 (TGF-[beta]1) and insulin-like growth factor II (IGF-II), has also been detected in JNA.

DIFFERENTIAL DIAGNOSIS:

1. Antrochoanal polyp
2. Nasopharyngeal carcinoma
3. Craniopharyngioma
4. Chordoma
5. Chondroblastoma
6. Rhabdomyosarcoma
7. Osteoma

BIOPSY

- Since the diagnosis is easily done and biopsy is dangerous, routine pre-op biopsy is not done.
- However Biopsy can be done in operating room under general anaesthesia in following circumstances(Batsakis)
 1. Female patients
 2. Ulcerative lesions
 3. Suspected rhabdomyosarcoma

RADIOLOGICAL INVESTIGATIONS

Computerised tomography helps in accurate preoperative assessment of the extent of the disease and aids in surgical planning and also in postoperative surveillance.

MRI scan is a very helpful tool in assessing the intracranial extension of JNA and also MR angiography aids in identifying the feeding vessel in a non invasive manner.

The tumour typically shows a “vascular blush” in the capillary phase which is typical of JNA.

HOLMAN MILLER SIGN

Forward bowing of the posterior wall of maxilla due to mass in the pterygopalatine fossa noted in the CT scan axial and sagittal view.

HANDOUSA SIGN

Maxillo-mandibular space widening due to extension of the mass from infratemporal fossa into the cheek.

Materials and methods

The present study consists of a series of '32' histopathologically proven cases of JNA. Some of the cases had been operated already for the disease and presented with recurrence.

This study was done at the Upgraded institute of otorhinolaryngology, Madras medical college, Chennai-03 during the period of May 2005 to Nov 2007.

Detailed history of each patient was taken. History of previous surgery and thorough clinical examination was done.

Diagnostic nasal endoscopy was done in all cases.

Following provisional diagnosis , all the cases were subjected to CT scanning with contrast. Selected cases also underwent MRI,MRA.

Angiography was done in cases with intracranial extension where blood supply is shared between in internal and external carotid arterial systems is expected and in cases planned for pre-op embolisation.

A Proforma was prepared for the study to record the history, clinical examination and investigations.

Grading of the tumour was done using Radowski staging system and appropriate surgical approach was employed.

Cases were studied for completeness of tumour removal, complications, blood loss, recurrence rate and morbidity.

Inclusion criteria

- Only advanced cases(Stage II a and above) were included in the study.
- Only cases histopathologically proved postoperatively were included in the study.

Exclusion criteria

- Cases with small tumours(Stage I) were excluded from the study.
- Cases with histopathology other than JNA or doubtful HPE were excluded.

Study period and centre

- May 2005 to November 2007.
- Upgraded institute of Otorhinolaryngology,MMC

Postoperative follow up

- Diagnostic nasal endoscopy in all patients every month for 6 months and once every 3 months thereafter.
- CT scan was done in patients suspected to have recurrence.

PROFORMA

Name :

Age :

Age at diagnosis :

Sex :

Presenting complaints:

1.Nasal bleed - yes/No

- Duration/no of episodes.

- amount of blood loss.

- Sponataneous/Provoked

- Releiving factors

2.Nasal obstruction

3. Headache

4.Facial pain

5. Change of voice- rhinolalia

6. Ocular complaints-double vision,Prominence of
eye ball ,dimness of vision.

History of previous surgery

Personal history

Family history

Examination

General examination	Pulse rate	Blood pressure
	pallor Yes/no	CVS/RS
	Surgical scars	

ENT examination

NOSE

External contour

Nasomaxillary groove

Septal position

Bleeding

Mass in nasal cavity

POST NASAL EXAMINATION :

- mass in nasopharynx
- bleeding
- size of mass, prominent blood vessels over the mass
- whether the mass occludes the Eustachian orifice.

EAR

Right

Left

Pinna

External ear

Tympanic membrane

ORAL CAVITY

- Palate
- Bulge present /or not
 - mobility of soft palate
 - scar

Eye

Proptosis

Vision

Ocular movement

Lacrimation

Lid closure

Face

Scar (if any)

INVESTIGATIONS

- Blood grouping and typing
- Complete hemogram
- Diagnostic nasal endoscopy

- Radiological

1. *CT scan paranasal sinuses*
 - plain and contrast
 - Axial and coronal views
 - in bone window.
 - 3 mm cuts
2. MRI and MR angiogram

SURGERY PERFORMED

1. Approach
2. Intraoperative findings
3. No of units of blood transfused
4. Embolisation Yes No Complication(if any)
5. External carotid ligation Yes No Complication(if any)

Histopathologic report

POSTOPERATIVE ANALYSIS

<i>Patient symptoms:</i>	Yes	No
1. Pain		
2. Swelling		
3. Redness		
4. Heat		
5. Loss of function		
6. Numbness		
7. Tingling		
8. Itching		
9. Rash		
10. Fever		
11. Chills		
12. Fatigue		
13. Headache		
14. Dizziness		
15. Nausea		
16. Vomiting		
17. Diarrhea		
18. Constipation		
19. Dry mouth		
20. Dry eyes		
21. Dry skin		
22. Dry hair		
23. Dry nails		
24. Dry throat		
25. Dry cough		
26. Dry wheezing		
27. Dry shortness of breath		
28. Dry chest pain		
29. Dry palpitations		
30. Dry irregular heart rate		
31. Dry fainting		
32. Dry dizziness		
33. Dry headache		
34. Dry nausea		
35. Dry vomiting		
36. Dry diarrhea		
37. Dry constipation		
38. Dry dry mouth		
39. Dry dry eyes		
40. Dry dry skin		
41. Dry dry hair		
42. Dry dry nails		
43. Dry dry throat		
44. Dry dry cough		
45. Dry dry wheezing		
46. Dry dry shortness of breath		
47. Dry dry chest pain		
48. Dry dry palpitations		
49. Dry dry irregular heart rate		
50. Dry dry fainting		
51. Dry dry dizziness		
52. Dry dry headache		
53. Dry dry nausea		
54. Dry dry vomiting		
55. Dry dry diarrhea		
56. Dry dry constipation		
57. Dry dry dry mouth		
58. Dry dry dry eyes		
59. Dry dry dry skin		
60. Dry dry dry hair		
61. Dry dry dry nails		
62. Dry dry dry throat		
63. Dry dry dry cough		
64. Dry dry dry wheezing		
65. Dry dry dry shortness of breath		
66. Dry dry dry chest pain		
67. Dry dry dry palpitations		
68. Dry dry dry irregular heart rate		
69. Dry dry dry fainting		
70. Dry dry dry dizziness		
71. Dry dry dry headache		
72. Dry dry dry nausea		
73. Dry dry dry vomiting		
74. Dry dry dry diarrhea		
75. Dry dry dry constipation		
76. Dry dry dry dry mouth		
77. Dry dry dry dry eyes		
78. Dry dry dry dry skin		
79. Dry dry dry dry hair		
80. Dry dry dry dry nails		
81. Dry dry dry dry throat		
82. Dry dry dry dry cough		
83. Dry dry dry dry wheezing		
84. Dry dry dry dry shortness of breath		
85. Dry dry dry dry chest pain		
86. Dry dry dry dry palpitations		
87. Dry dry dry dry irregular heart rate		
88. Dry dry dry dry fainting		
89. Dry dry dry dry dizziness		
90. Dry dry dry dry headache		
91. Dry dry dry dry nausea		
92. Dry dry dry dry vomiting		
93. Dry dry dry dry diarrhea		
94. Dry dry dry dry constipation		
95. Dry dry dry dry dry mouth		
96. Dry dry dry dry dry eyes		
97. Dry dry dry dry dry skin		
98. Dry dry dry dry dry hair		
99. Dry dry dry dry dry nails		
100. Dry dry dry dry dry throat		
101. Dry dry dry dry dry cough		
102. Dry dry dry dry dry wheezing		
103. Dry dry dry dry dry shortness of breath		
104. Dry dry dry dry dry chest pain		
105. Dry dry dry dry dry palpitations		
106. Dry dry dry dry dry irregular heart rate		
107. Dry dry dry dry dry fainting		
108. Dry dry dry dry dry dizziness		
109. Dry dry dry dry dry headache		
110. Dry dry dry dry dry nausea		
111. Dry dry dry dry dry vomiting		
112. Dry dry dry dry dry diarrhea		
113. Dry dry dry dry dry constipation		
114. Dry dry dry dry dry dry mouth		
115. Dry dry dry dry dry dry eyes		
116. Dry dry dry dry dry dry skin		
117. Dry dry dry dry dry dry hair		
118. Dry dry dry dry dry dry nails		
119. Dry dry dry dry dry dry throat		
120. Dry dry dry dry dry dry cough		
121. Dry dry dry dry dry dry wheezing		
122. Dry dry dry dry dry dry shortness of breath		
123. Dry dry dry dry dry dry chest pain		
124. Dry dry dry dry dry dry palpitations		
125. Dry dry dry dry dry dry irregular heart rate		
126. Dry dry dry dry dry dry fainting		
127. Dry dry dry dry dry dry dizziness		
128. Dry dry dry dry dry dry headache		
129. Dry dry dry dry dry dry nausea		
130. Dry dry dry dry dry dry vomiting		
131. Dry dry dry dry dry dry diarrhea		
1		

1. Palatal defect/nasal regurgitation
2. Nasal obstruction

3. Epistaxis
4. Watering of eyes
5. Anosmia
6. Numbness over the face
7. External Facial deformity

No of recurrences:

Time of recurrence after surgery:

Grade of recurrent tumour:

Clinical examination:

Nose-

Ear –

Throat-

Diagnostic nasal endoscopy:

CT scan:

SURGICAL PROCEDURE

In this study , Endoscopic transnasal approach was used in 23 cases.

Maxillary swing procedure was done in 4 cases.

Transpalatine approach, previously the commonest approach employed was used only in three cases.

Combined transantral and transpalatine excision was done in one case.

Combined Transpalatine and endoscopic excision was done in one case.

Transantral excision was done in two cases.

Lateral rhinotomy was employed in one case.

All the cases were done under Hypotensive

anaesthesia(Halothane,Propofol,Nitroglycerine drip)

External carotid artery ligation was done in 8 cases.

Embolisation was done in 5 cases.

Amount of Blood transfused :-

Minimum – no transfusion.

Maximum – 4 units.

Recurrent tumour was seen in 8 cases. Recurrent tumour was commonly seen in the pterygopalatine fossa.

Postoperative post nasal packing was done in all cases and were removed on second postoperative day in the operating room.

All cases were postoperatively treated with Inj.Cefotaxim 1gm i.v B.D, Inj.Metronidazole 500 mg i.v B.D for 7 to 10 days.

Postoperative follow up was done every month for six months and every 6 months thereafter. It consists of diagnostic nasal endoscopy and CT if any suspicion of recurrence.

10 cases were lost to follow up after 5 months.

SURGICAL PROCEDURES DONE IN OUR STUDY

In this study Endoscopic Transnasal approach was the commonest approach employed.

Performed under general anaesthesia. Throat packing was done in all cases.

Endoscopic transnasal approach :

Position of the patient- supine with 15 deg head end elevation.

Nose is gently packed with cottonoids soaked in 4% xylocaine and 1 in 10,000 adrenaline solution,15 minutes prior to the surgery.

The nasal cavity is first visualized with 0 degree Karl storz endoscope. Local infiltration is given over the root of middle turbinate, uncinate process, nasal septum,posterior end of middle turbinate using 1in 100,000 adrenaline and 1% xylocaine mixture.

The procedure starts with uncinectomy,partial middle turbinectomy and a wide middle meatal antrostomy.Complete ethmoidectomy and sphenoidotomy were done.

The posterior wall of maxillary sinus,often pushed forwards and thinned out in cases of JNA is removed and the Pterygopalatine fossa is exposed.

The attachments of the mass are easy to delineate due to the excellent direct visualization offered by endoscope. Sphenopalatine artery is cauterized.The attachments are systematically released using monopolar cautery.

The extensions into the pterygopalatine fossa and infratemporal fossa are removed using 45 deg and 70 deg endoscopes and angled instruments.

After releasing from its attachments the mass is pushed posteriorly into the nasopharynx.

Doyens mouth gag is applied and the softpalate is retracted using suction catheters passed through the opposite nostril.

The mass is now delivered in toto through the oral cavity.Any remnants can be visualized,cauterized and removed under direct vision through the nose.

Postnasal packing is done using either 14 size Foleys catheter or conventional postnasal packs. Anterior nasal packing is done if anterior oozing is still present. Wide spectrum intravenous antibiotics are used for 3 – 4 days.

Post nasal packs are removed on postop day 2 in the operating room ,keeping things ready for a repack. The patient is discharged on 4 th or 5 th postop day.

MAXILLARY SWING APPROACH

This is relatively new procedure described by wei and associates for approach to nasopharyngeal tumours. We have used this approach quite often in our institute for excision of extensive stage II c /III lesions where endoscopic approach cannot be used.

Wide spectrum antibiotic coverage is started on the day of surgery. The procedure is done under general anaesthesia. Temporary tarsoraphy is done. After an initial external carotid artery ligation on the side of the lesion, a Weber- Ferguson- Longmire Facial incision is made. The incision extends 0.5 cm below the lower lid margin and is extended laterally till the preauricular region. The incision curves around the medial canthus and extends along the nasomaxillary groove and wind around the nasolabial sulcus to reach the philtrum. Now a lip splitting incision is made and the incision is extended along the

gingivobuccal sulcus turned around the last molar tooth and comes forward between the two hard palates in the midline.

Flaps are raised and contrary to maxillectomy the anterior maxillary wall is left attached to the masseter flap. Osteotomies are made just below the infra orbital rim, through the floor of the ipsilateral nasal cavity, and through the posterior wall of the maxillary sinus. The pterygoid plates are then separated from the maxillary tuberosity with a osteotome.

The maxilla, which is left pedicled to the masseter and cheek flap can now be swung out of the way to gain wide exposure of the nasopharynx and parapharyngeal space.

Now the entire tumour with all its extension can be seen. The mass is separated from its attachments by blunt finger dissection. The mass is delivered in toto. Hemostasis is secured by placing hot saline pads in the cavity.

The maxilla is repositioned and fixed by wiring along the lines of osteotomies. The skin is sutured with atraumatic 3-0 ethilon sutures. Ryles tube is inserted. Oral feeds are started from second postop day. Sutures are removed on the sixth post op day. Patient is discharged after 8 to 10 days.

TRANSPALATINE APPROACH

Anaesthesia – GA with orotracheal intubation, tracheostomy.

Position – Supine ,Rose position.

Boyle Davis mouth gag is applied .Care is taken to see that the blade of the instrument does not occlude the anaesthetic tube or damage the posterior pharyngeal wall by pressure.

The mucosa over the palate is infiltrated with 2 % xylocaine and 1:100000 adrenaline 5 minutes before making the incision. A Curved incision , bowed forwards is made with a 15 size bard parker blade between the two maxillary tuberosities,keeping internal to the greater palatine foramen.

Adequate exposure to the postnasal space can be obtained through an insision that extends just in front of the posterior margin of the hard palate.

Incision is made through the mucosa and periosteum down to the bone. Inorder to simplify subsequent suturing, it is easy to start by elevating for a short distance, the edge of the mucoperiosteal flap on the anterior side of the incision. The posterior flap is then separated completely from the undersurface of the hard palate.

A small right angled dissector is used to separate the flap from the posterior margin of the bone. But a knife may be needed to free the attachment to the posterior spine of the hard palate.

Mucosa on the upper surface of the palate is divided transversely and the postnasal space is examined.

Near the pterygo mandibular raphe and the pterygoid hamulus , tensor palati muscle fans out from the tendon passing round the hamulus and if necessary it is divided just medial to the hamulus. Soft palate is retracted by means of two rubber catheters placed in the retracted site. Care is taken not to jeopardize the greater palatine blood supply to the soft palate. By retracting the entire tumour is visualized. The mass is then freed from its attachments by blunt dissection.

Then by using a periosteal elevator, mass is mobilised from sphenopalatine foramen region. The mass is then held with a babcock forceps and the entire mass is removed in toto by side by side movement along with continuous lift by perisoteal elevator.

Cavity is packed with hot saline pad. After achieving hemostasis, the flaps are sutured in the midline. Vicryl is used and the sutures are allowed to absorb by themselves.

Ryles tube is inserted and oral feeds are started by second post op day. The patient is discharged 8 to 10 days after the surgery.

TRANS ANTRAL APPROACH

A temporary tarsoraphy is done . Weber – Ferguson incision is made about 2-3 mm below the lid margin along the lower lid. This allows the skin to be separated from the underlying orbicularis oculi and minimize the postoperative edema.

Incision curves inferiorly just lateral to the medial canthus and extends along the nasomaxillary groove and the curves around the nasolabial fold to reach the columella. Then a lip splitting incision is made and the incision is carried along the gingivo buccal sulcus upto the maxillary tuberosity.

And the palatine of the incision is not done. The anterior wall of the maxilla removed by osteotomy. Posterior wall of the maxilla is removed. Maxillary artery is identified and ligated.

The mass can be visualized and released from its attachments and removed in one piece. Hemostasis secured with hot saline packs. Wound closed in layers. Cavity is packed with antibiotic smeared gauze and postnasal packing done.

LATERAL RHINOTOMY (MOURE' s)

The upper part of the incision need to be no higher than the point midway between the medial canthus and dorsum of the nose. To avoid an unsightly depressed scar it should run just medial to the nasomaxillary groove but follow the ala to finish within the ipsilateral nasal cavity.

All layers are divided including the nasal mucosa and retraction of the alar region away from the incision is done. Periosteum and skin is elevated over the nasal bone and the fronto nasal process of maxilla ,so as to allow full visualization of the mass.

Medial maxillectomy is done and the mass is removed in toto after separating from the attachments. Anterior and posterior nasal packing is done. Wound is closed in layers.

COMBINED TRANSPALATINE AND TRANS ANTRAL APPROACH

This approach was done in one of the cases in our study. It was done since the tumour was extensive. But this approach does not result in any better exposure than medial maxillectomy and resulted in more morbidity and prolonged hospital stay.

COMBINED TRANSPALATINE AND ENDOSCOPIC APPROACH

Used in one of the cases in our study, the procedure was started as an endoscopic approach and after releasing the lateral attachments, transpalatine approach was used to release attachments to the nasopharynx.

OTHER APPROACHES

SARDANA 'S APPROACH

Sublabial incision is combined with transpalatine approach to facilitate removal of lateral extensions of the mass. Sublabial incision is made from the midline, extending upto the maxillary tuberosity.

Surgeon 's index finger is inserted into pterygoid space and blunt dissection is used to free the tumour from its lateral attachments. It can then be delivered into the nasopharynx.

FACIAL TRANSLOCATION

This procedure is done by the ENT surgeon jointly with the neuro surgeon. A extended Weber- Ferguson incision is made and a inferiorly based flap is raised.

Cranio facial skeleton is exposed. Osteotomies are performed to free the anterior face of the maxilla, the malar eminence,zygomatic arch,inferior and lateral orbital rims and the orbital floor. Orbito-maxillary complex is removed and preserved for later replacement.

An osteotomy is performed at the base of the coronoid process and temporalis muscle is transposed inferiorly. A Fronto temporal

osteotomy is then performed and the foramen ovale , spinosum and rotundum as well as the superior orbital fissure are identified.

Surgical field can extend from the contralateral Eustachian tube to the ipsilateral geniculate ganglion and from the superior orbital fissure and cavernous sinus area to the level of the hard palate. It included the nasopharynx ,clivus, sphenoid,cavernous sinus and infratemporal fossa.

After tumour removal dura is repaired and temporalis muscle is mobilized to line the defect. Orbito maxillary complex is then replaced and fixed. skin is sutured in layers.

MIDFACIAL DEGLOVING (Conley and Price)

Sublabial approach to the nasal and nasopharyngeal cavities in which the midface is essentially degloved from the anterior maxillary facial skeleton by extension of a sublabial incision bilaterally. This exposure provides generous access to sinuses and nasopharyngeal cavity.

The maxillary antrum as well as the pterygomaxillary spaces are readily accessible.

TRANSMANDIBULAR APPROACH (Kermen)

The incision begins vertically in front of the ear and carried down the neck anterior to the sternomastoid muscle. Dissection is started in the neck by exposing the carotid artery bifurcation at which level the external carotid is ligated.

Lower pole of parotid is dissected free and the vessels entering the substance from below are transected. At this level one severs the insertion of masseter muscle into the mandible. Incision is curved down to the periosteum at the angle of mandible so as to expose its entire lateral aspect. Periosteum is stripped upwards from the lateral aspect of the ascending ramus of the mandible.

Transection of the mandible is done with gigli saw at a point about 1 cm below the coronoid and condyloid processes. Separation and retraction of mandibular fragments expose tubomuscular wall of the nasopharynx which is incised on its lateral wall, so that its lumen is entered.

Tumour is exposed and dissected out. Nasopharynx is closed. Mandible is approximated with stainless steel wires.

DENKERS MODIFICATION

Tumour in maxillary antrum and anterior part of the nose may removed by sublabial incision as well as by performing a caldwel-luc antrostomy using Denker's modification of removing the lateral wall of the nose.

TRANSHYOID APPROACH

This is suitable for the tumour localized to nasopharynx without extension into the surrounding structure. The disadvantage is that usually requires temporary tracheostomy for two to three days.

TRIPLE APPROACH (HIRANANDANI)

Transpalatal and lateral rhinotomy approach is combined with caldwel incision. Complete exposure of pterygo palatine fossa is possible by removing posterior wall of maxillary antrum after opening the antrum through caldwel –Luc incision.

Once the pterygo palatine fossa is exposed, tumour can be easily palpated bimanually with one finger in fossa and another in the nasopharynx. The whole tumour is removed in toto under direct vision.

MODIFIED BARBOSA TECHNIQUE

Weber Ferguson incision extending from the eye to the tragus is made.

Three bony cuts are made , two at the zygoma and one at the coronoid process to reach the infratemoral fossa.

FISCH INFRATEMPORAL FOSSA APPROACH

TYPE- C

For extensive cases of JNA with intracranial extension. The morbidity of the procedure includes conductive hearing loss on the involved side and anaesthesia over the distribution of V2 and V3.

Endoscopic and KTP laser-assisted surgery

This is a recent modality of surgery where the endoscopic approach is combined with KTP laser assisted dissection to minimize bleeding and improve exposure and precision or tumour removal. Needs further evaluation before accepting as a standard modality of treatment.

HEMOSTATIC METHODS

External carotid artery ligation

Neck is extended by placing a sand bag under the shoulder and the head is partially rotated to the other side. Curved incision following one of the skin folds of the neck is made centered over the bifurcation of the common carotid artery at the upper border of the thyroid cartilage.

Incision is carried through the platysma muscle to the deep cervical fascia. The great auricular nerve should be preserved. Deep cervical fascia is divided along its attachment to the sternomastoid muscle and infrahyoid muscles are exposed by blunt dissection.

Common Facial vein is ligated and divided. Carotid sheath is opened to expose the common carotid artery and jugular vein with the vagus nerve lying posteriorly. Bifurcation of common carotid artery is at the level of the upper border of the thyroid cartilage.

Posterior belly of the digastric muscle and the stylohyoid muscle should be identified where they cross lateral to the internal and external carotid arteries and retracted to expose the carotid bulb. Hypoglossal nerve should be carefully identified where it crosses lateral to both the arteries.

External carotid artery is identified by finding one of its distal branches. Variations in the origin and arrangement of all the branches of the external carotid must be expected. External carotid artery is ligated with silk sutures. Deep cervical fascia is approximated to the sternomastoid muscle. Skin including the platysma is closed with interrupted silk sutures.

CAROTID ANGIOGRAM AND EMBOLISATION

Informed high risk consent is taken from the patient and his attendants. Groin is shaved .Nil oral for an appropriate period prior to procedure to avoid aspiration during a possible reaction to the contrast medium. Premedication with inj. Pentazocine(fortwin) 30 mg im/iv is given.

The procedure is done within 48- 72 hours prior to the surgery. After cleansing the area with a suitable preparation , 5 to 10 ml of 1- 2% xylocaine is infiltrated around the femoral artery.

Percutaneous needle puncture of an artery (femoral) is done. Inner stylet is removed when the needle is in the lumen of the artery and is replaced with guide wire of appropriate diameter. The needle is removed and a catheter or introducer is inserted over the wire.

Then the guide wire is withdrawn. Then injection of contrast through the catheter is done(IOHEXOL). Maxillary artery and feeding vessel of the tumour is identified by the tumour blush. Embolisation of the feeding vessel is done with gel foam beads.

After the procedure is over , the catheter is withdrawn and tight bandage is applied over the area. Wound site is checked at regular intervals. Patient should remain on bed rest overnight following the procedure.

COMPLICATIONS

1. Hemorrhage
2. vascular thrombosis
3. vascular stenosis
4. pseudo aneurysm formation
5. local sepsis
6. allergy to contrast
7. air embolism
8. vasovagal reaction
9. loss of guide wire
10. Cerebro vascular accident with paralysis- 1 case
11. Death due to embolisation of internal carotids- 1 case

Radiotherapy

Fitzpatrick recommendation is followed, wherein the patient is subjected to low dose radiotherapy of 3000 – 3500 rads over a period of 3 weeks. Doses higher than this do not improve the treatment outcome and adversely increase the chance of radiation induced malignancy in the patient.

Complications:

Mucositis, Osteoradionecrosis, pituitary necrosis and growth defects, radiation induced malignancy.

Chemotherapy

Chemotherapy is an alternative form of therapy

Unresectable tumors can be given chemotherapy for palliation

Adriamycin and decarbazine

Extensive regression of tumor

Possible alternative to radiation?

OBSERVATION

In this study of 32 locally advanced cases of Juvenile angiofibroma, the clinical presentation was as follows

Mass in nasal cavity, nasopharynx, Pterygopalatine fossa – 20 cases

Mass extending to infratemporal fossa - 8 cases

Extension to the cheek - 1 case

Extension to orbit - 1 case

Intracranial extension - 4 cases

Clinical Presentation

1. Nasal obstruction – all cases
2. Epistaxis - 31 cases
3. Cheek involvement - 1 case
4. Proptosis - 1 case

One of our patient presented only with history of nasal obstruction and had no history of epistaxis. The diagnosis was done based on clinical appearance of the tumour and CT findings.

Treatment Modality

Surgery - 29 cases

1. Endoscopic excision -23 cases
2. Maxillary swing - 4 cases
3. Transpalatal excision - 3 cases
4. Combined approach - 2 cases
5. Other approaches - 3 cases

Radiotherapy- 2 cases

STAGING OF DISEASE

Stage II A – 4 cases (12.5%)

Stage II B – 16 cases (50%)

Stage II C – 8 cases (25%)

Stage III A – 3 cases (9.4%)

Stage III B – 1cases (3.1%)

DISCUSSION

In our case study , 32 cases of Juvenile nasopharyngeal angiofibroma were managed by surgery and radiotherapy. Hormonal therapy was not tried in any of our cases. In all the cases, history was evaluated carefully.

Of the 32 cases 18 cases were primary cases and 14 cases were recurrent cases.

The average age of presentation of primary cases was 14 to 16 yrs. The youngest was 9 yrs and the oldest was 29 yrs old. The average age of presentation of recurrent cases was 18 to 21 yrs. The youngest was 9 yrs and the oldest was 31 yrs.

Nasal obstruction and epistaxis were the commonest symptoms. Anosmia, Rhinolalia, cheek swelling and proptosis were the other symptoms. The average delay in diagnosis was 4 months.

Preoperative Computerised tomographic scanning in coronal and axial views were done in all cases. Magnetic resonance imaging and MR angiography were done in 7 cases. The extent of the tumour was assessed accurately in all cases. Staging of the disease with Radowsky staging system based on CT and MRI findings was done. Only stage II a and above cases were included in the study.

Carotid angiography with embolisation was done in 5 cases. The internal maxillary artery was the commonest feeding vessel. We had one post embolisation death and one case had quadriparesis following the procedure, which recovered with conservative management.

Various hemostatic methods were used in our study. External carotid artery ligation was done in 8 cases. It is a safe and effective procedure and decreases the vascularity of the tumour considerably.

Embolisation helps in decreasing the vascularity of the mass and also shrinks the mass. It was done in 5 cases . We had two embolisation related complications. Embolisation results in shrinkage of tumour leading to incomplete resection.Hence we prefer external carotid ligation over embolisation in our cases.

Additionally Hypotensive anaesthesia was used in all cases. Pre op Tab. Diazepam 5 mg , Tab. Atenolol 25 mg were administered the night before surgery. Propofol, halothane and nitroglycerine drip were used intraoperatively for hypotensive anaesthesia.

Diagnostic nasal endoscopy is a very useful and economic tool for follow up of the cases and CT scan can be used to evaluate patients suspected to have recurrences. With DNE recurrences can be diagnosed very early.

RADOWSKI STAGING SYSTEM FOR JNA

(Radkowski et al,16 1996)

Stage I - Tumour limited to nose, nasopharynx, sinuses.

I A- tumour limited to nasal cavity and or nasopharyngeal vault.

I B- tumour extending to > one sinus.

Stage II -

II A – Minimal occupation of the PMF

II B – full occupation of the PMF

II C – extension posterior to the pterygoid plates

Stage III

III A – Erosion of the skull base with minimal intracranial
extension.

III B – Skull base erosion with extensive intracranial
extension with or without cavernous sinus involvement.

In this study , twenty three patients underwent endoscopic excision of the tumour making it the commonest approach employed in our study. Of these patients the stage distribution was as follows,

Stage II A – 1 case

Stage II B – 13 cases

Stage II C – 4 cases

Stage III A – nil

Stage III B – nil

Recurrent cases – 5 cases(All were Stage II)

Maxillary swing approach and removal was done in 4 cases. Two cases were stage II C and two cases were stage III A.

Transpalatine surgery, previously the most common surgical route for JNA excision was done only in 3 cases. All cases were Stage II A.

Combined Transpalatine and endoscopic excision was done in one case (Stage II B).

Combined transantral and transpalatine approach was done in one case.

(Stage II B).

Transantral excision was done in two cases (stage II B- 1 case, Stage II C- 1 case)

Lateral rhinotomy was done in one case (Stage II B)

Radiotherapy was given in two cases.(Stage II B)

Posterior nasal packing was done in all the cases. The pack was removed on postoperative day II in the operating room .

Intraoperative external carotid artery ligation was done in 8 cases.

Embolisation of the feeding vessel was done in 5 cases.Of these patients one died of massive Cerebrovascular accident. One patient had Quadripareisis which improved with conservative management.

Mass was sent for Histopathological examination in all cases for confirmation of diagnosis. All the cases was treated with intravenous antibiotics (Cefotaxim and Metronidazole).

There were seven cases of recurrence. All the cases were taken up for surgery. Cause of the recurrence was due to persistence of mass in the pterygopalatine fossa.

Of these seven cases of recurrence, six cases underwent endoscopic excision and one case underwent maxillary swing procedure.

Palatal fistula was seen in three cases. This was managed conservatively in two cases and one case had local advancement flap closure.

Lacrimal sac injury and dacryocystitis was seen in 4 cases. Two cases underwent endoscopic dacryocystorhinostomy.

Synechiae with chronic sinusitis was seen 5 cases and one case underwent functional endoscopic sinus surgery and all other case were managed conservatively.

CONCLUSION

In this study , 32 cases were analysed. Nasal obstruction and epistaxis were the common symptoms noticed in almost all cases.

CT scan was done in all cases . MRI and MRA was done in 7 cases. These investigations were used to accurately assess the extent of the tumour and plan appropriate surgical procedure. Staging based on Radowski ‘ s system, a widely accepted and comprehensive staging system was employed. Intracranial spread was noted in four cases.

Carotid angiogram with embolisation was done in five cases. The typical capillary phase tumour blush was seen in all cases. The internal maxillary artery was the feeding vessel in all cases. One patient died following the procedure and another patient had quadriparesis.

External carotid artery ligation in our opinion is a safe and effective method for decreasing blood loss during surgery. Combined with hypotensive anaesthesia provides a very good blood less field, essential for complete tumour removal.

The likelihood of death due to JNA is very low. Therefore, the efficacy of a particular treatment is evaluated based on local control and complications. Transnasal endoscopic excision is ideal procedure for JNA excision. The advantages of this approach are listed below,

1. Tumour removal under direct magnified vision
2. Least morbidity
3. Decreased hospital stay
4. Decreased blood loss
5. Can be repeated any number of times in cases of recurrences with no extra morbidity.
6. Decreased operating time

Among the external approaches for removal of extensive tumour of grades more than II C, the maxillary swing is an excellent approach for removal of such extensive tumours. It gives good exposure and less morbidity compared to a transantral or maxillectomy approach.

Radiotherapy can be given for cases with extensive intracranial extensions where surgical removal would result in unacceptable morbidity. The local control rates

approach 73-100 %. Any recurrences after radiotherapy can be managed by salvage surgery.

The complications encountered in this study were,

1. recurrence
2. Palatal perforation
3. Lacrimal sac injury
4. Sinusitis

Recurrences can be minimized by proper pre operative evaluation of the CT and MRI scan and planning the appropriate management approach.

Palatal perforation are usually seen following transpalatal approach and can be managed conservatively with an palatal obturator. Refractive cases can be managed surgically with flap closure.

Lacrimal sac injury occur in maxillary swing, lateral rhinotomy, transantral approach and can be managed by an endoscopic Dacryocystorhinostomy.

Lasers are recent addition in the management tools for JNA excision. Its role and effectiveness needs to be evaluated.

The likelihood of cure after surgery is approximately 80% to 85%; the probability of ultimate local control approaches 100%.

The vast majority of patients have lesions that are completely resectable with relatively limited morbidity and should be treated surgically. A small subset of patients present with recurrent and/or extensive tumours with significant intracranial invasion where the likelihood of complete resection with acceptable morbidity is modest. These patients are best treated with moderate-dose RT.

Bibiliography

- Martin H, Ehrlich HE, Abels JC. Juvenile nasopharyngeal angiofibroma. *Ann Surg* 1948;127:513-536.
- Bremer JW, Neel HB III, De Santo LW, et al. Angiofibroma: Treatment trends in 150 patients during 40 years. *Laryngoscope* 1986; 96: 1321-1329.
- Cansiz H, Guvenc MG, Sekecioglu N. Surgical approaches to juvenile nasopharyngeal angiofibroma. *J Craniomaxillofac Surg*. 2006 Jan;34(1):3-8. Epub 2005 Dec 15.
- Cummings BJ, Blend R, Keane T, et al. Primary radiation therapy for juvenile nasopharyngeal angiofibroma. *Laryngoscope* 1984; 94: 1599-1605.
- Douglas R, Wormald PJ. Endoscopic surgery for juvenile nasopharyngeal angiofibroma: where are the limits? *Curr Opin Otolaryngol Head Neck Surg*. 2006 Feb;14(1):1-5.
- Enepekides DJ. Recent advances in the treatment of juvenile angiofibroma. *Curr Opin Otolaryngol Head Neck Surg*. 2004 Dec;12(6):495-499.
- Hardillo JA, Vander Velden LA, Knecht PP. Denker operation is an effective surgical approach in managing juvenile nasopharyngeal angiofibroma. *Ann Otol Rhinol Laryngol*. 2004 Dec;113(12):946-950.

- Herman F, Lot G, Chapot R, et al. Long term follow up of juvenile nasopharyngeal angiofibromas: Analysis of recurrences. Laryngoscope 1999; 109: 140-147.

- Hosseini SM, Borghei P, Borghei SH, Ashtiani MT, Shirkhoda A.
Angiofibroma: an outcome review of conventional surgical approaches. Eur Arch Otorhinolaryngol. 2005 Oct;262(10):807-812. Epub 2005 Mar 1.

- Andrews JC, Fisch U, Valavanis A, Aeppli U, Makek MS. The surgical management of extensive nasopharyngeal angiofibromas with the infratemporal fossa approach. Laryngoscope 1989;99: 429-437.

- Roger G, Tran Ba Huy P, Froelich P, et al. Exclusively endoscopic removal of juvenile nasopharyngeal angiofibroma: trends and limits. Arch Otolaryngol Head Neck Surg 2002;128:928-935.

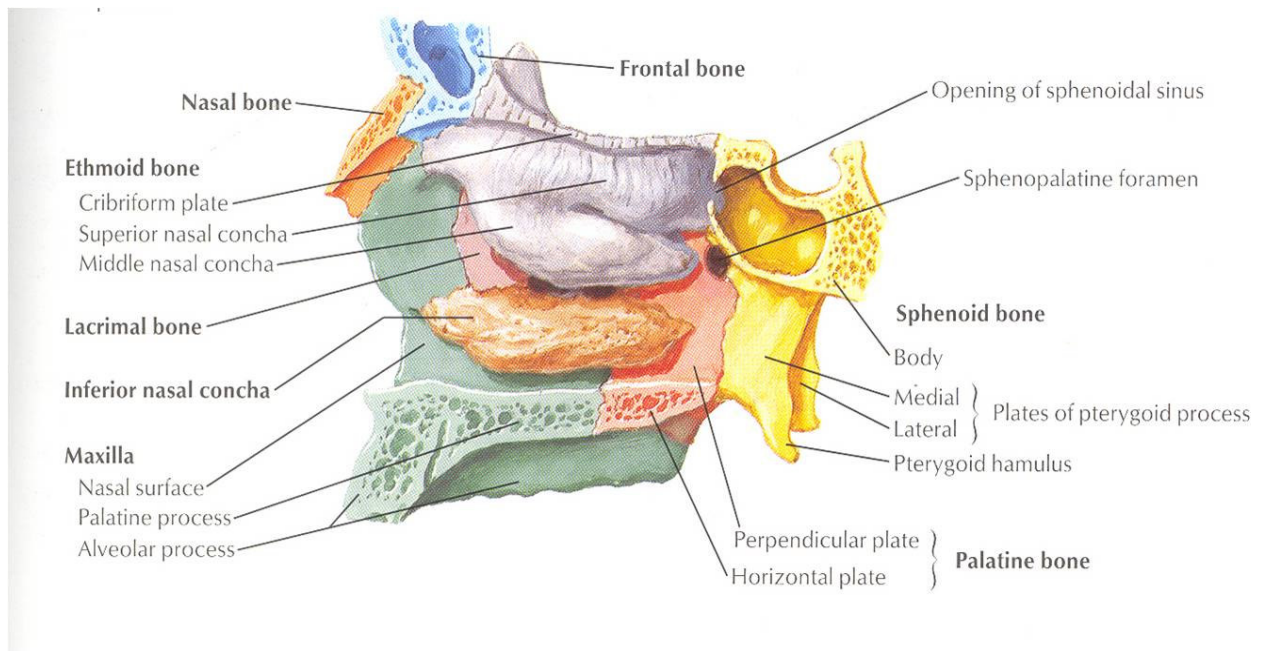
- Hagen R, Romalo G, Schwab B, Hoppe F, Schweikert HU. Juvenile nasopharyngeal fibroma: androgen receptors and their significance for tumor growth. Laryngoscope 1994;104:1125-1129.

- Deschler DG, Kaplan MJ, Boles R. Treatment of large juvenile

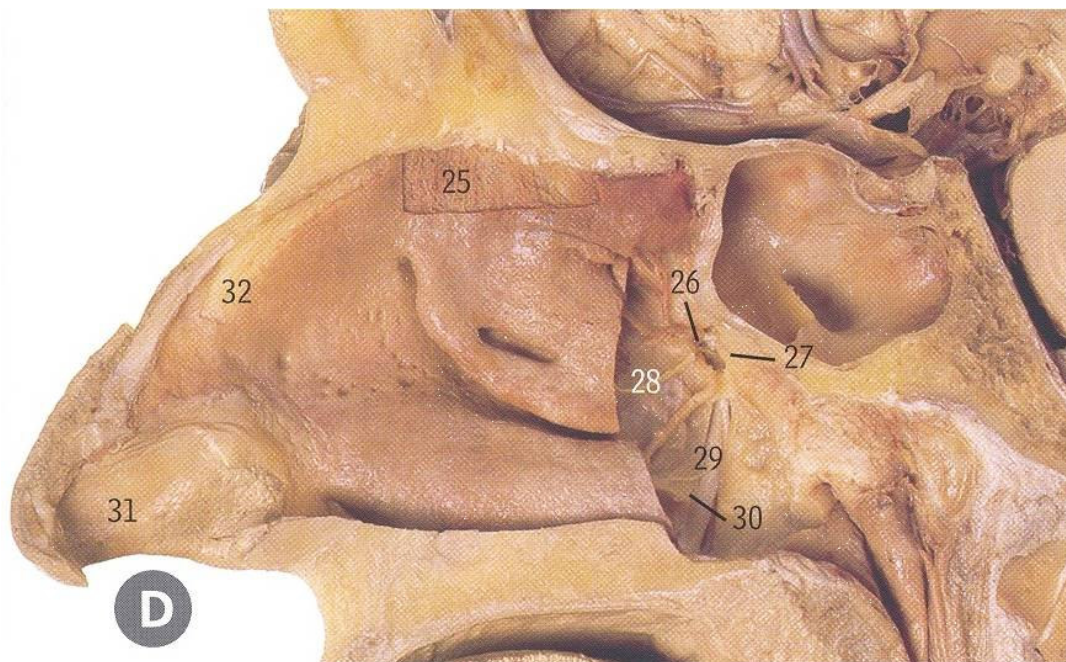
nasopharyngeal angiofibroma. Otolaryngol Head Neck Surg
1992;106:278-284

■ Radkowski D, McGill T, Healy GB, Ohlms L, Jones DT. Angiofibroma.
Changes in staging and treatment. Arch Otolaryngol Head Neck Surg
1996;122:122-129

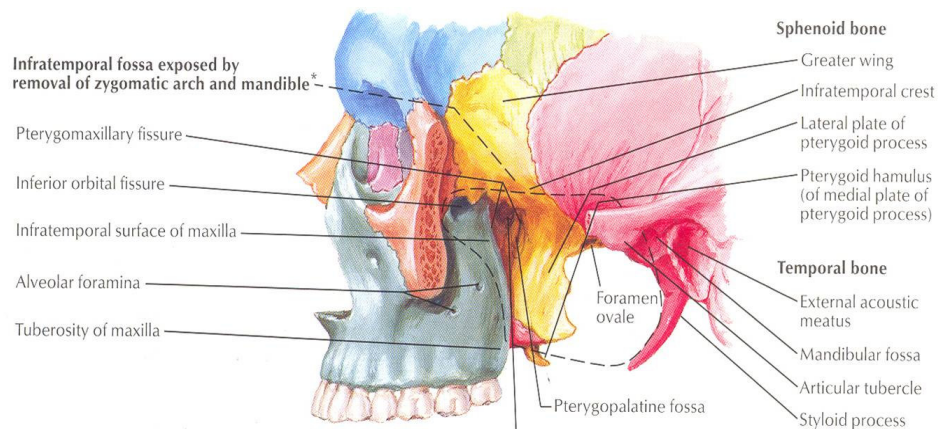
■ Frequent b-Catenin Mutations in Juvenile
Nasopharyngeal Angiofibromas
Susan C. Abraham, Elizabeth A. Montgomery,
Francis M. Giardiello, and Tsung-Teh Wu
American Journal of Pathology, Vol. 158, No. 3, March 2001
Copyright © American Society for Investigative Pathology



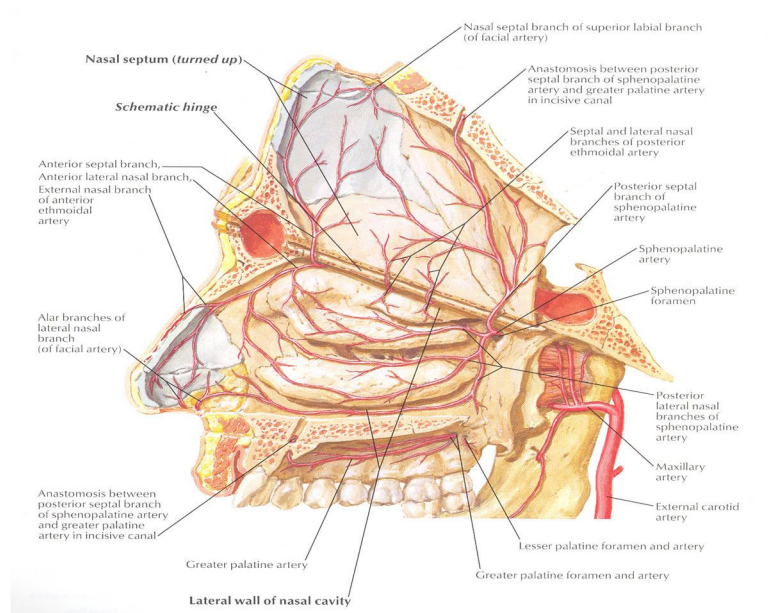
ANATOMY OF LATERAL NASAL WALL AND SPHENOPALATINE FORAMEN



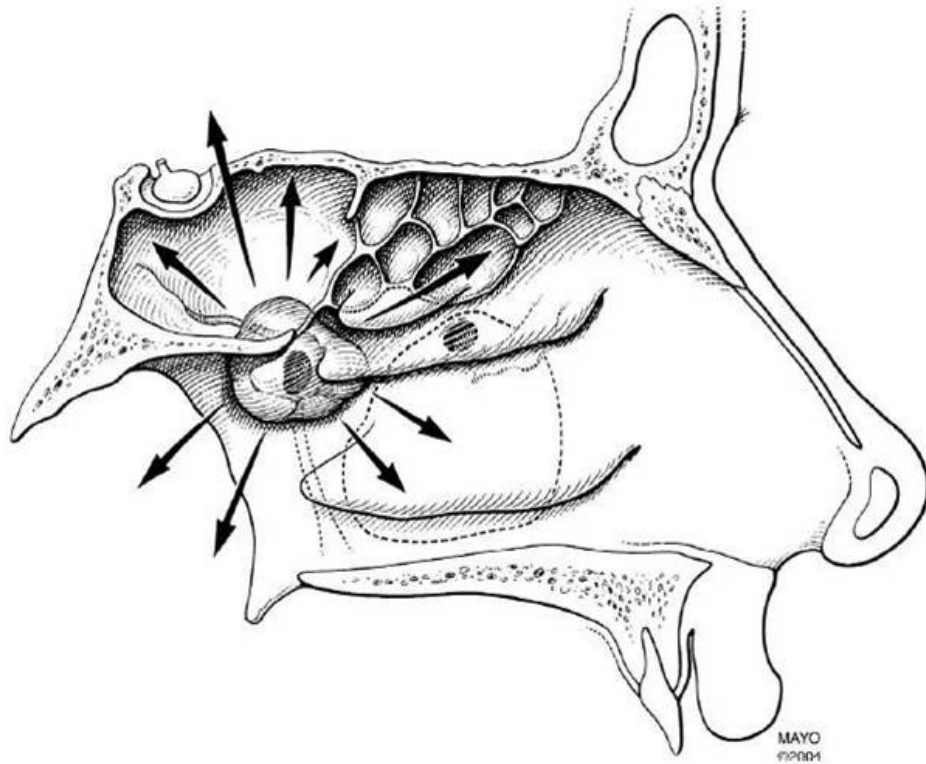
SPHENOPALATINE FORAMEN WITH ITS CONTENTS



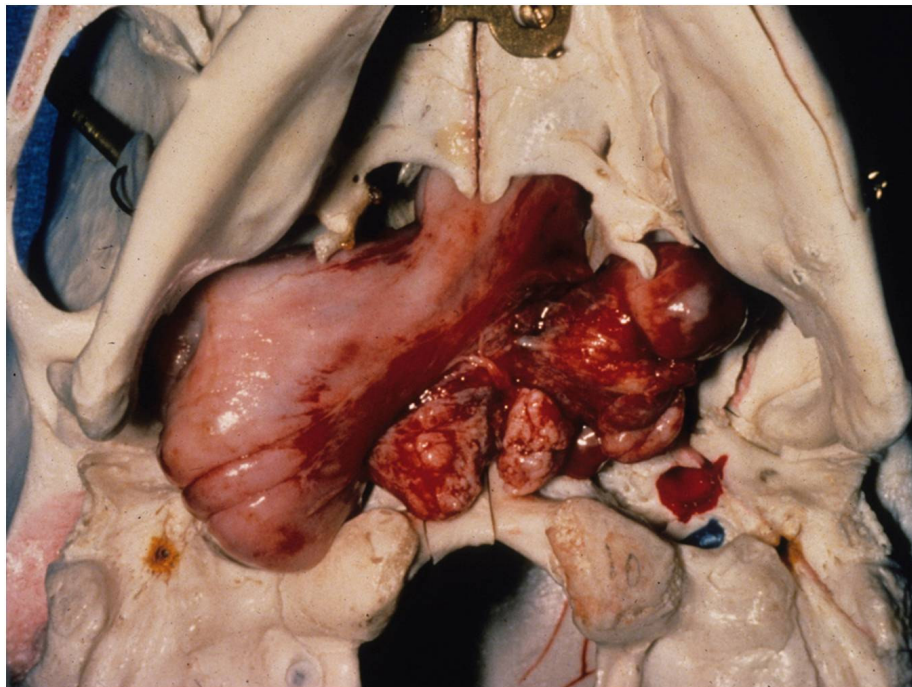
ANATOMY OF INFRATEMPORAL FOSSA



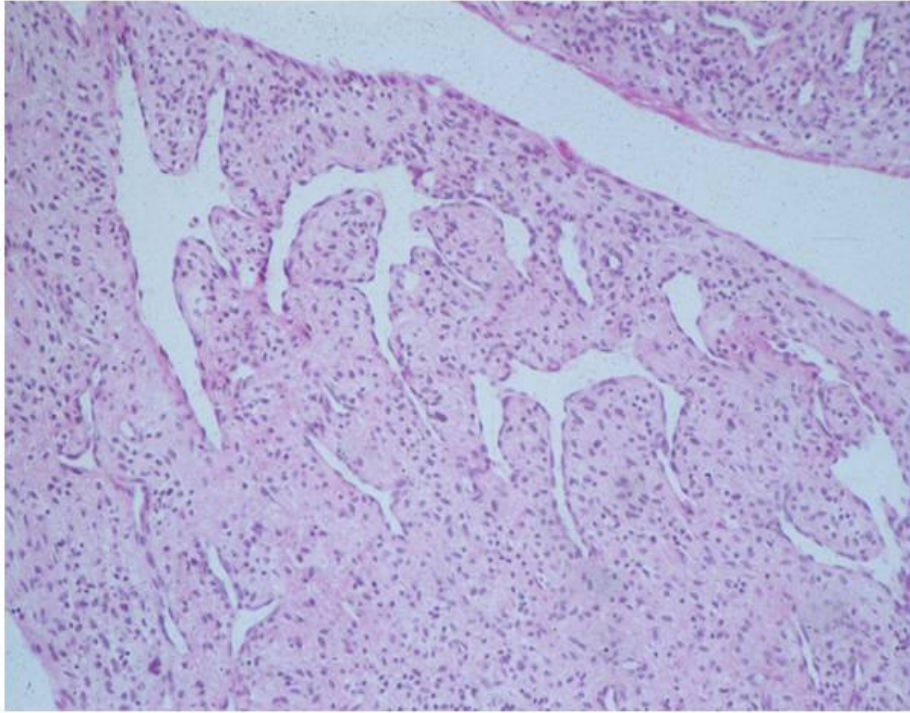
BLOOD SUPPLY OF THE NASAL CAVITY



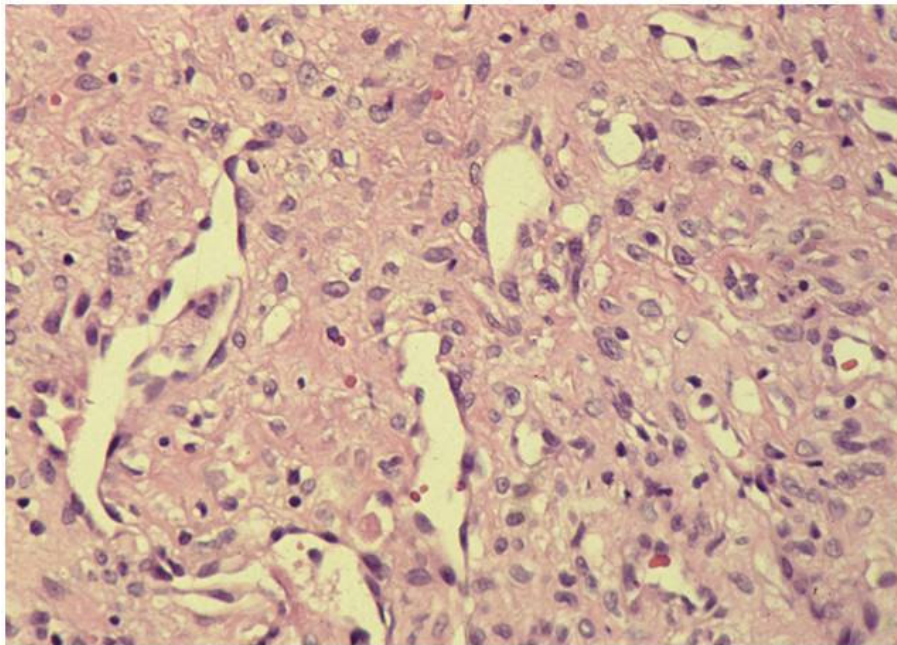
ROUTES OF SPREAD



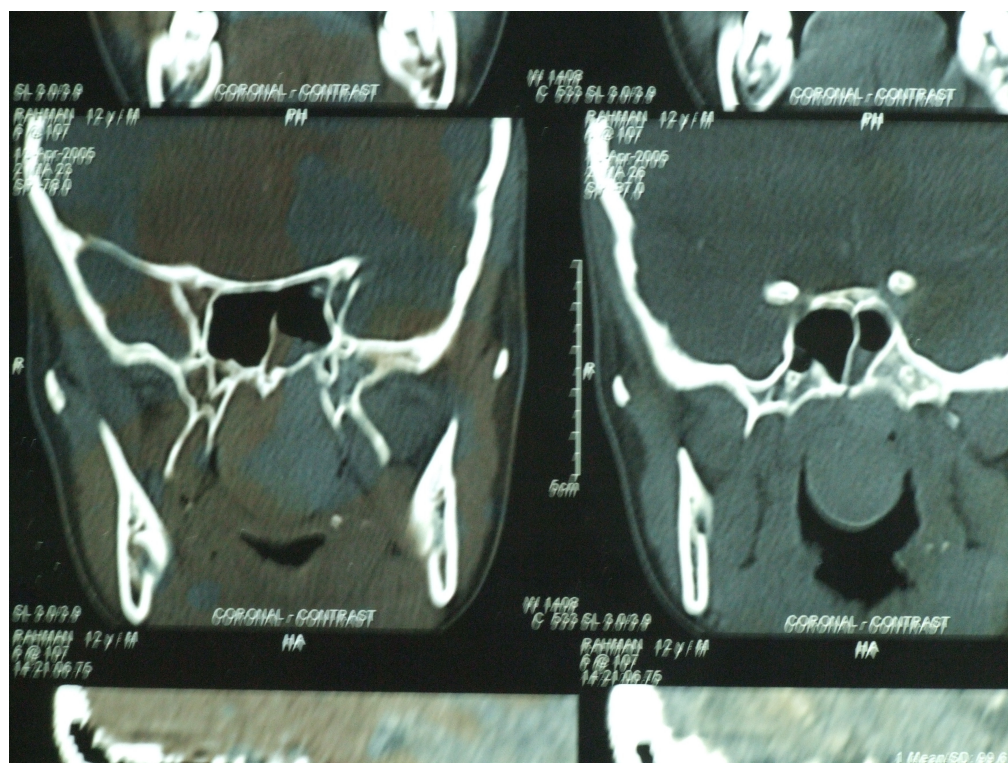
EXTENSIONS OF THE TUMOUR DEPICTED ON A SKULL



LOW POWER EOSIN HEMATOXYLIN STAIN



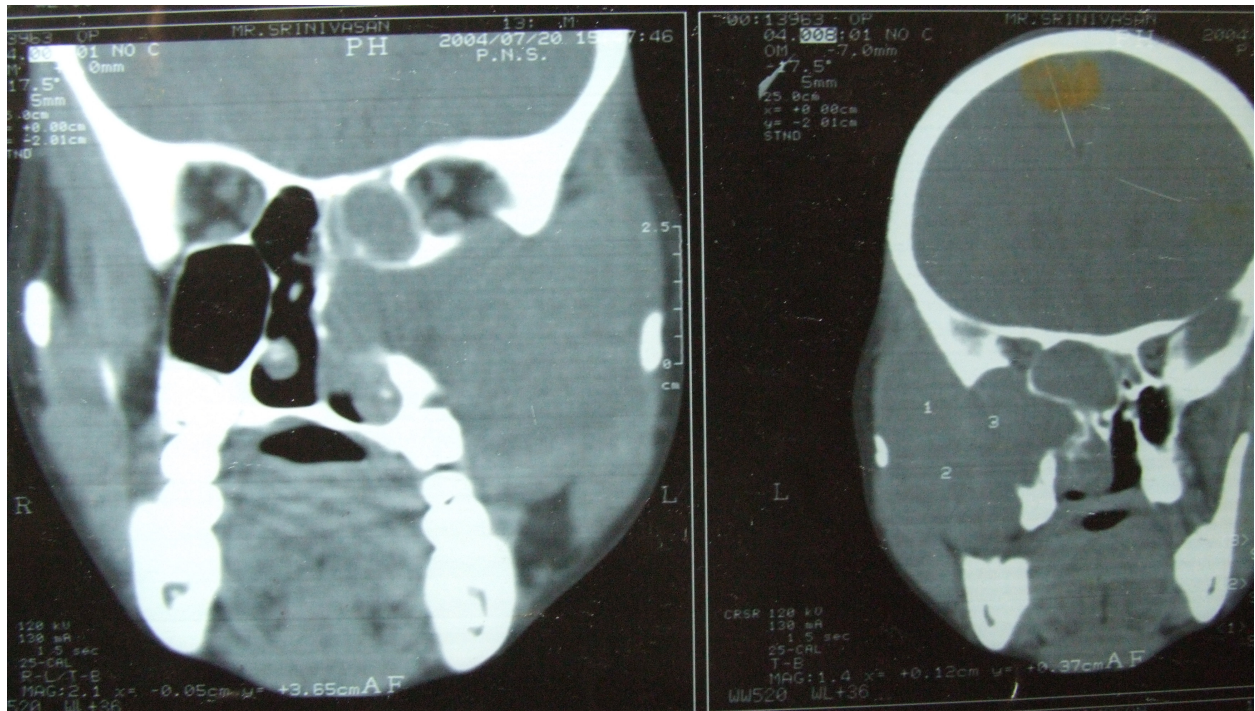
HIGH POWER VIEW OF THE SAME SLIDE



CT SCAN CORONAL VIEW- STAGE II a LESION



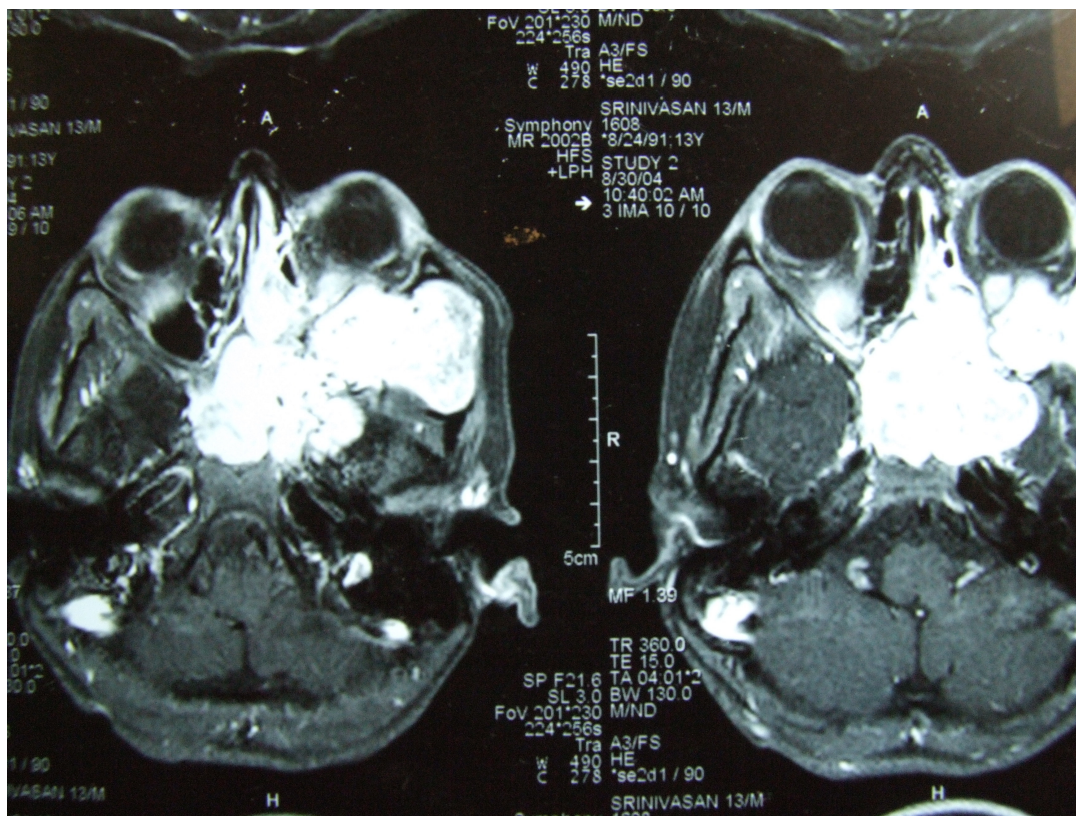
CT SCAN CORONAL VIEW – STAGE II b LESION



CT SCAN CORONAL VIEW – STAGE II c LESION



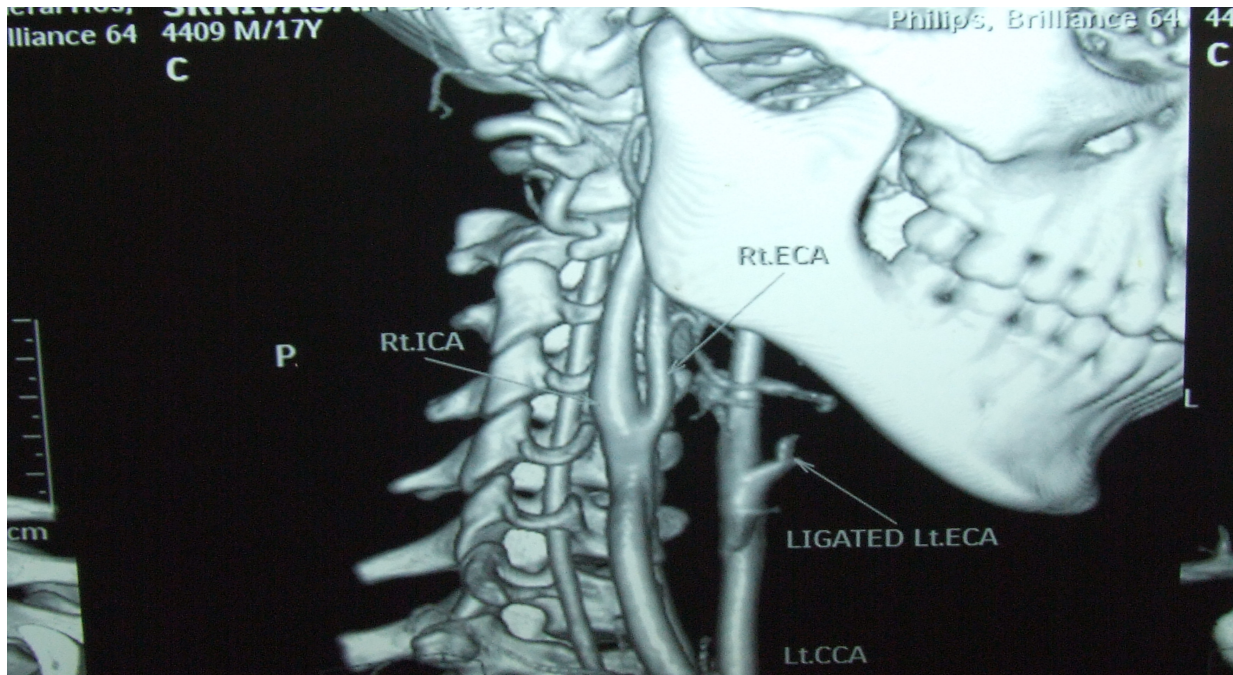
CT SCAN SAGGITAL VIEW- STAGE III a LESION WITH INFERIOR ORBITAL FISSURE EXTENSION



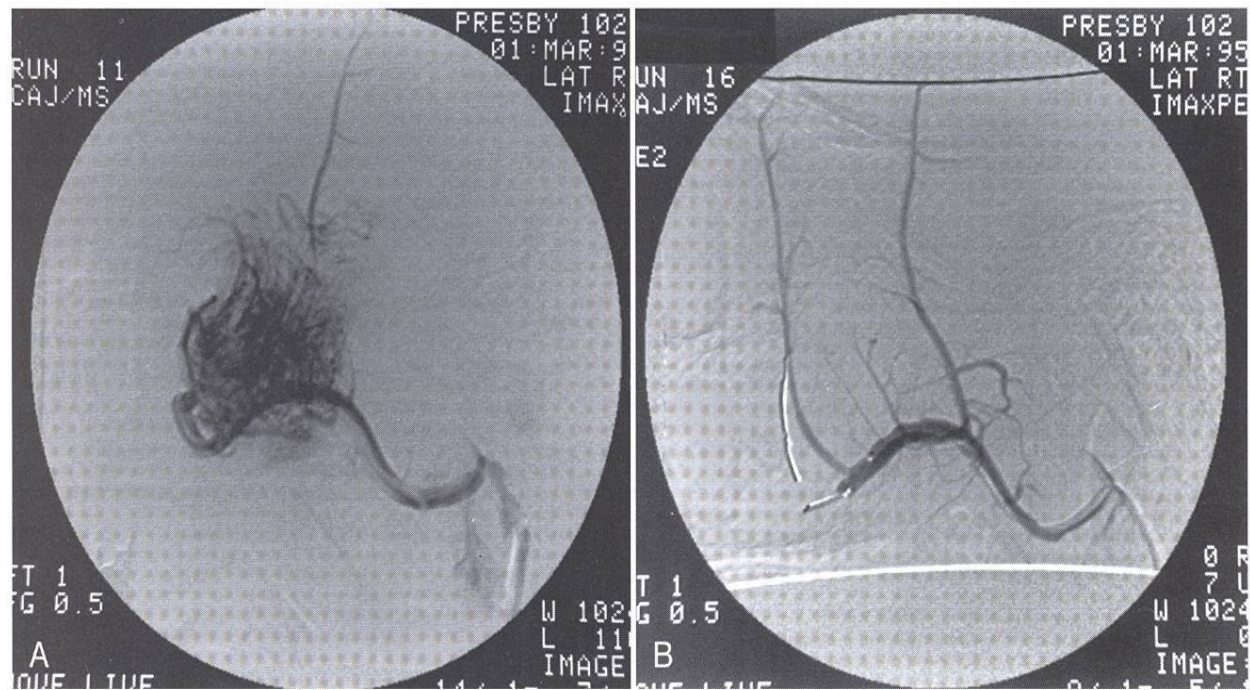
MRI AXIAL VIEW -STAGE II c LESION (NOTE HYPERVASCULARITY)



CT SCAN SAGGITAL VIEW – ANTRAL SIGN



64 SLICE SCANNING SHOWING LIGATED ECA



CAROTID ANGIOGRAM WITH EMBOLISATION



PATIENT AFTER ENDOSCOPIC EXCISION



PATIENT AFTER EXTERNAL PROCEDURE



PALATAL FISTULA FOLOWING MAXILLARY SWING PROCEDURE

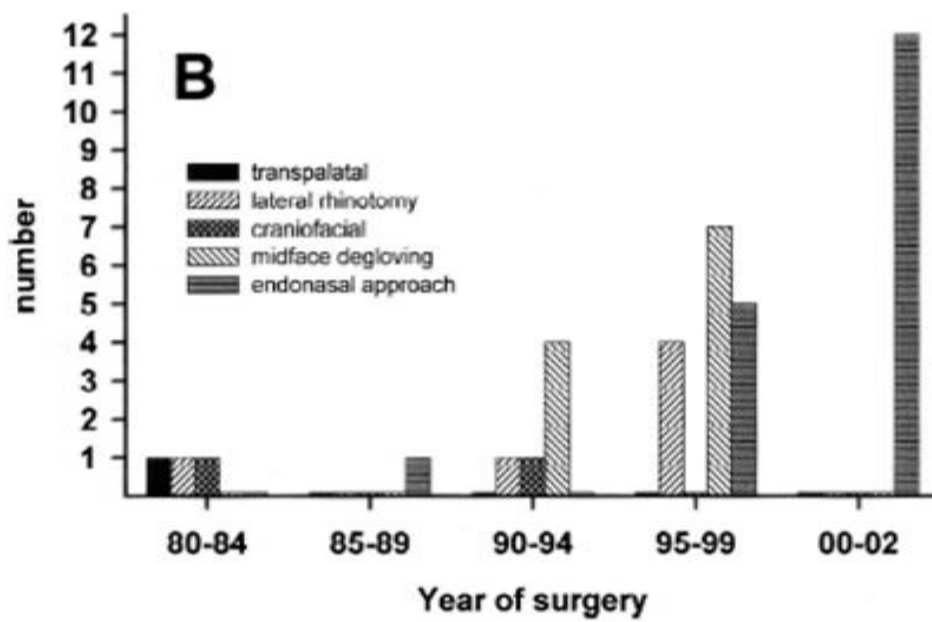
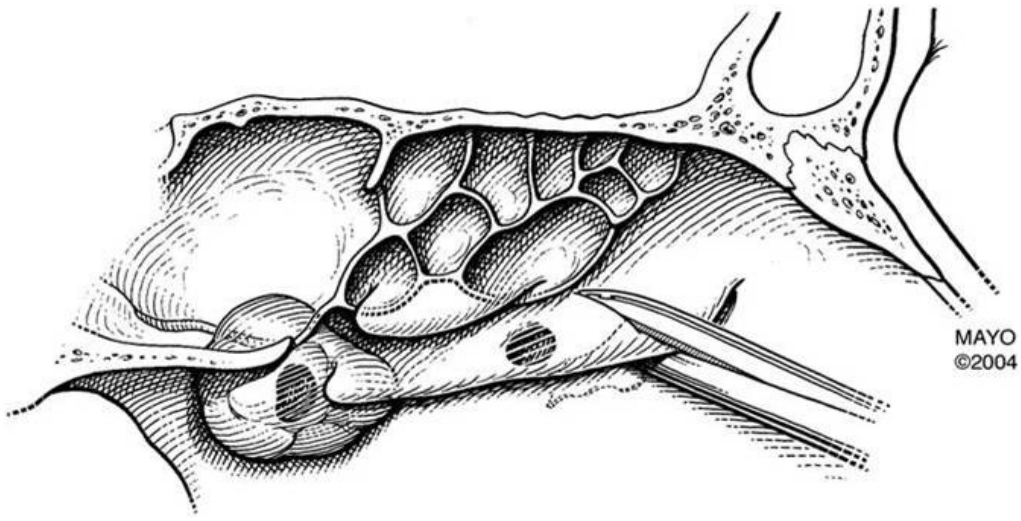
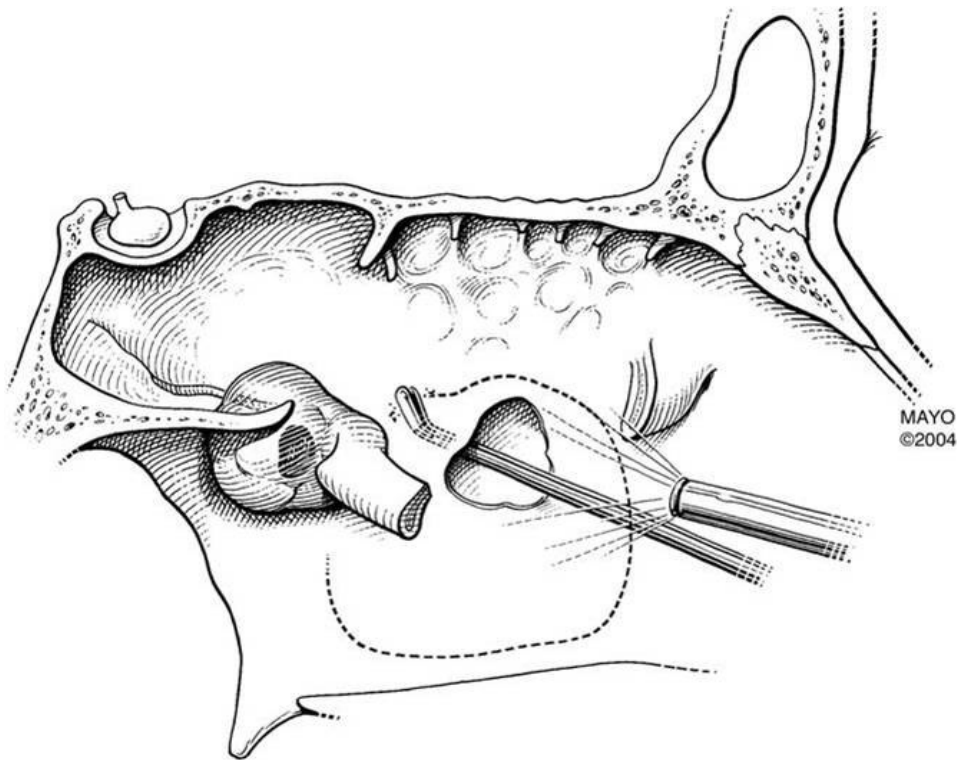


CHART SHOWING CHANGE IN SURGICAL APPROACHES

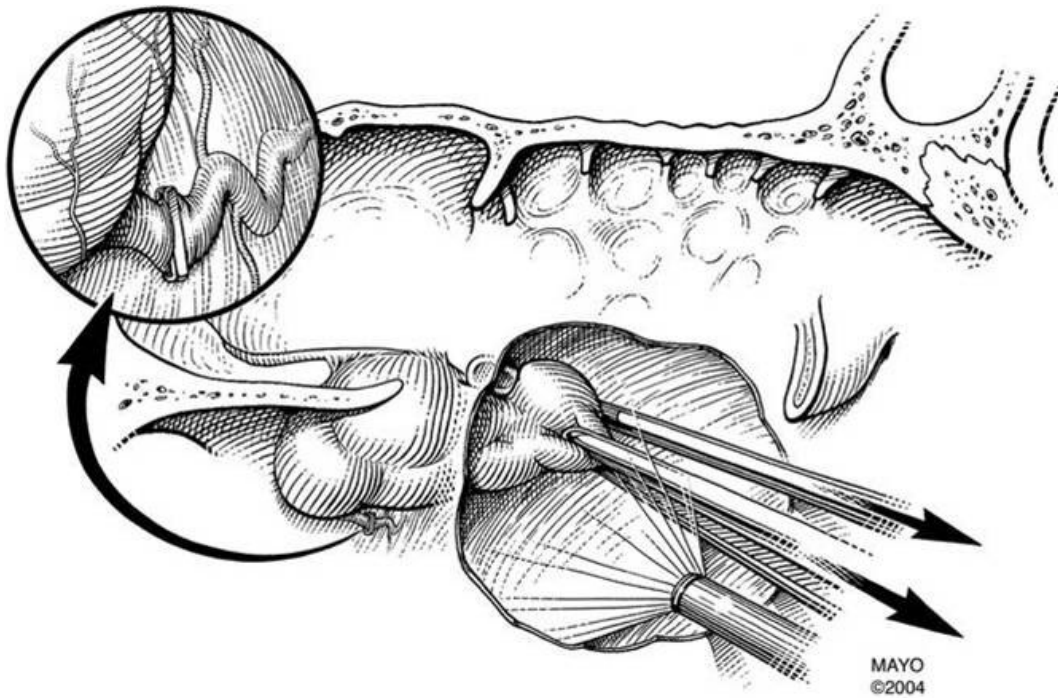
ENDOSCOPIC EXCISION



PARTIAL MIDDLE TURBINECTOMY AND TUMOUR LOCATION

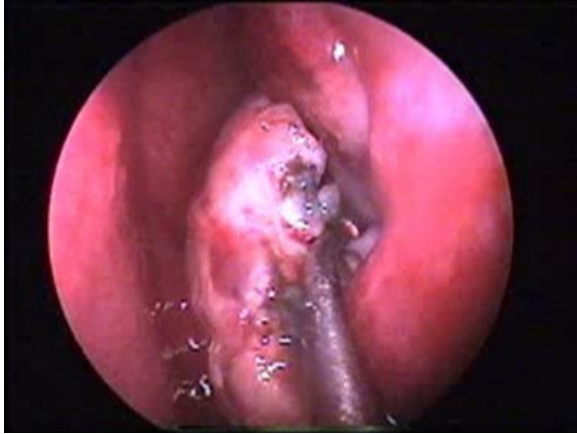


OPENING OF POSTERIOR WALL OF MAXILLA TO EXPOSE SPHENOPALATINE FORAMEN

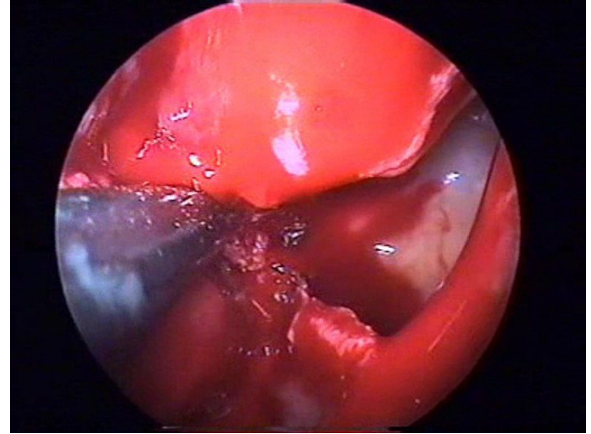


TUMOUR REMOVAL UNDER DIRECT VISION
LIGATION OF FEEDING VESSEL

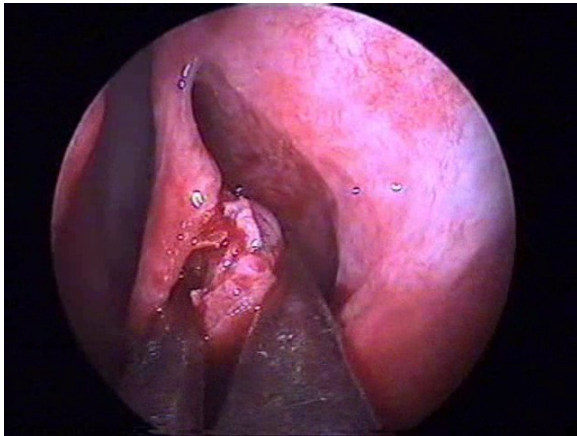
ENDOSCOPIC EXCISION



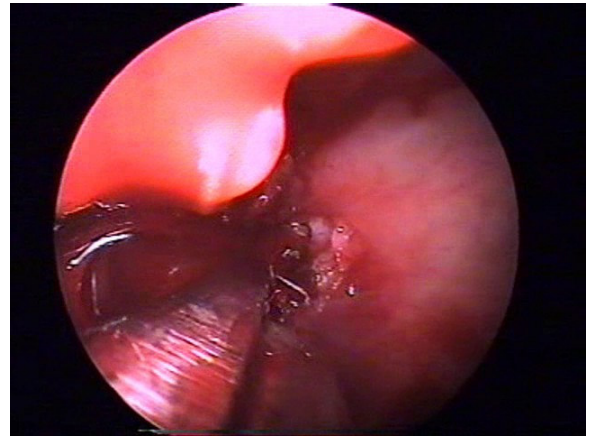
TUMOUR
APPEARANCE



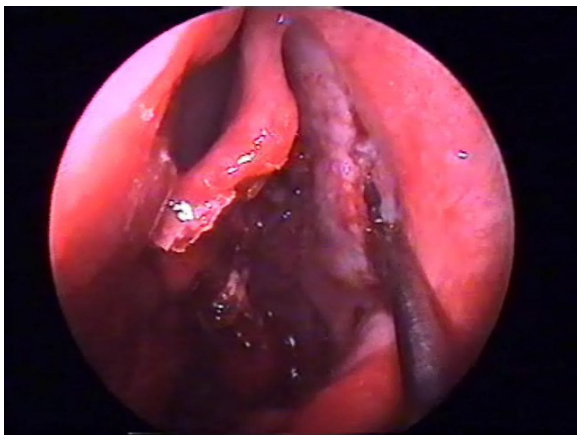
WIDE MIDDLE MEATAL
ANTROSTOMY



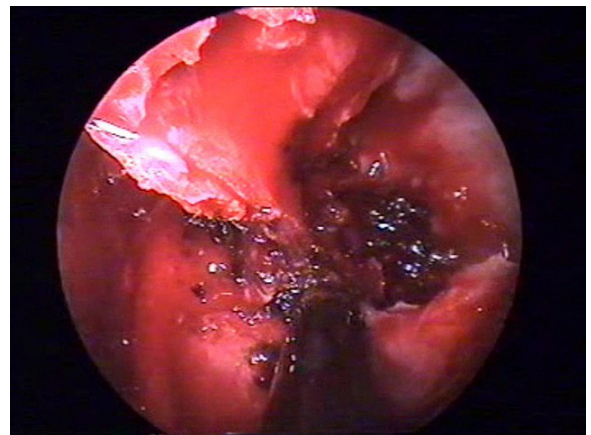
PARTIAL MIDDLE TURBINATE
RESECTION



REMOVAL OF POSTERIOR WALL
OF MAXILLA



UNCINECTOMY



CAUTERY OF FEEDING VESSEL

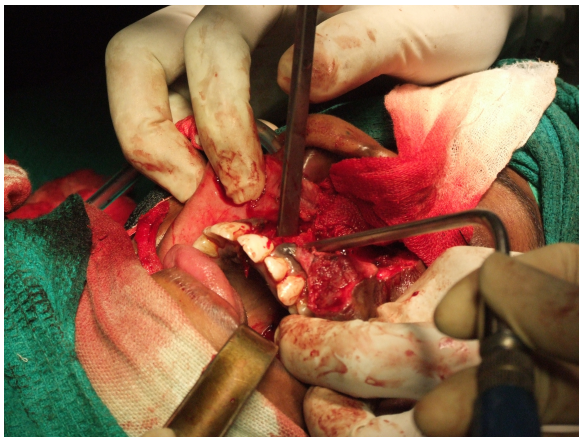
MAXILLARY SWING PROCEDURE



INCISION



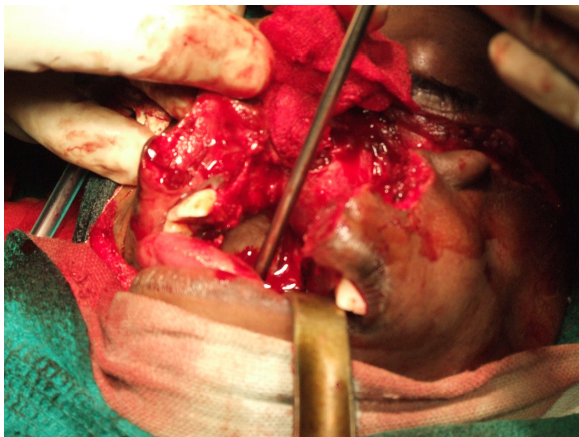
TUMOUR



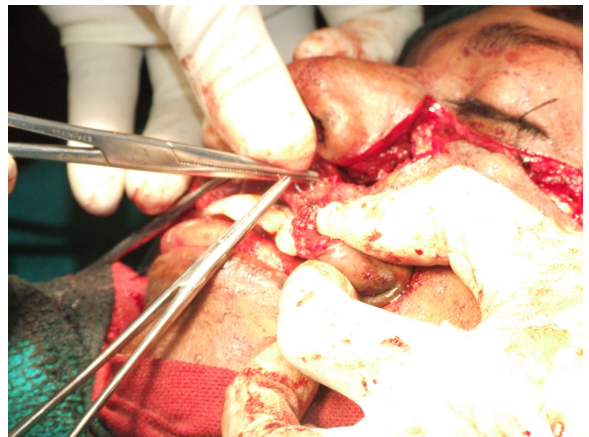
OSTEOTOMY



AFTER TUMOUR REMOVAL



FLAP RAISAL



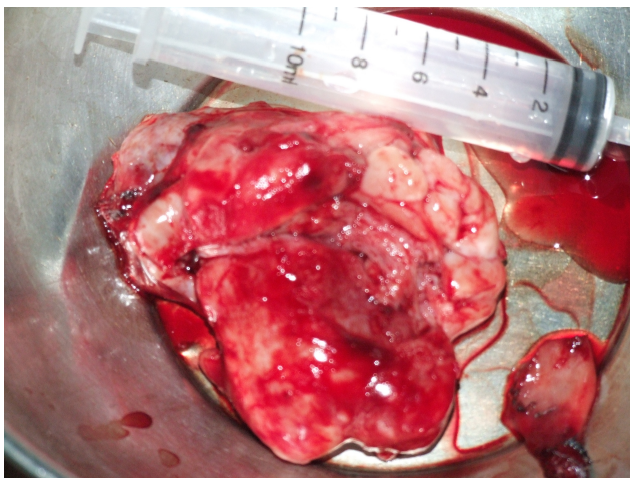
ENDOSCOPIC EXCISION

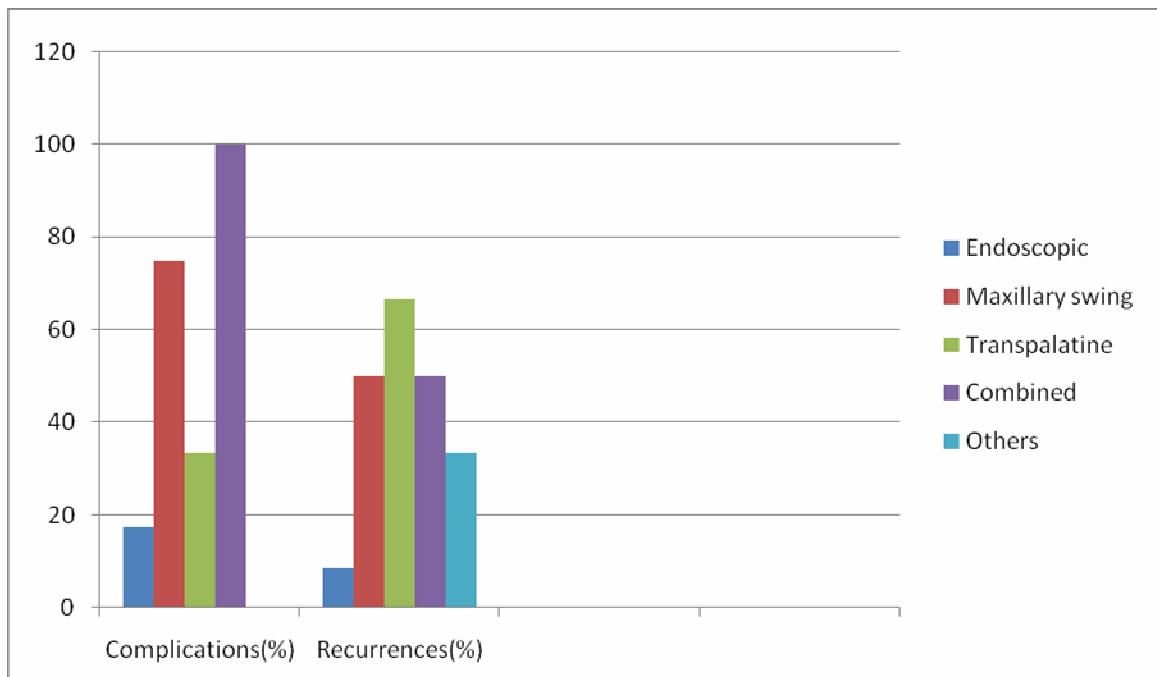
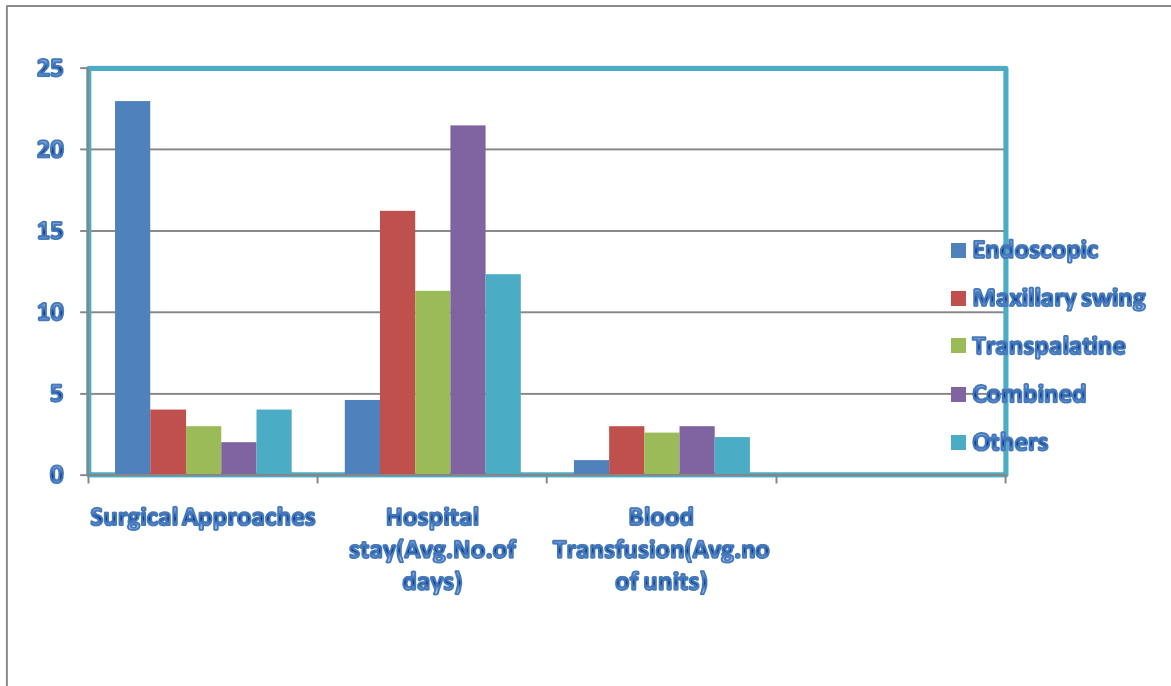
MAXILLARY SWING PROCEDURE



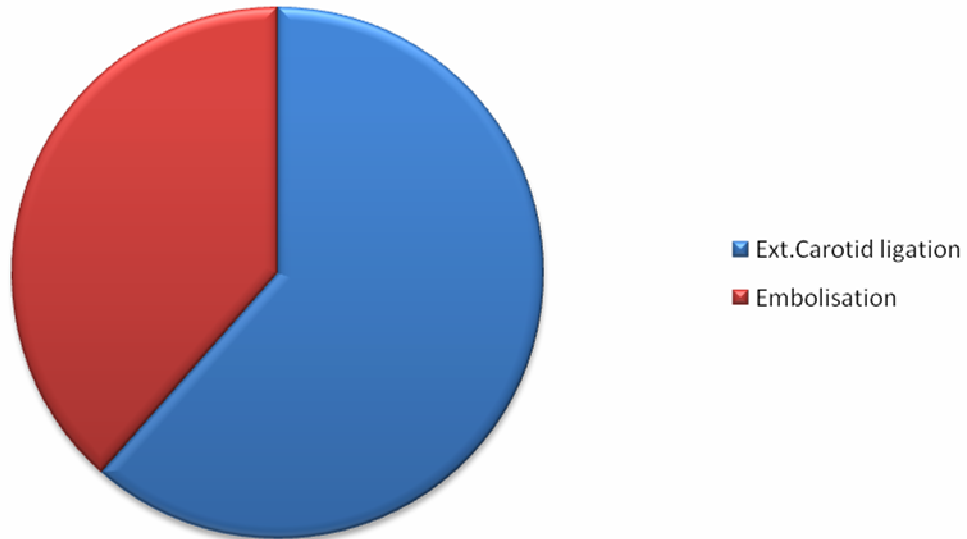
AFTER WOUND CLOSURE

TUMOUR SPECIMEN

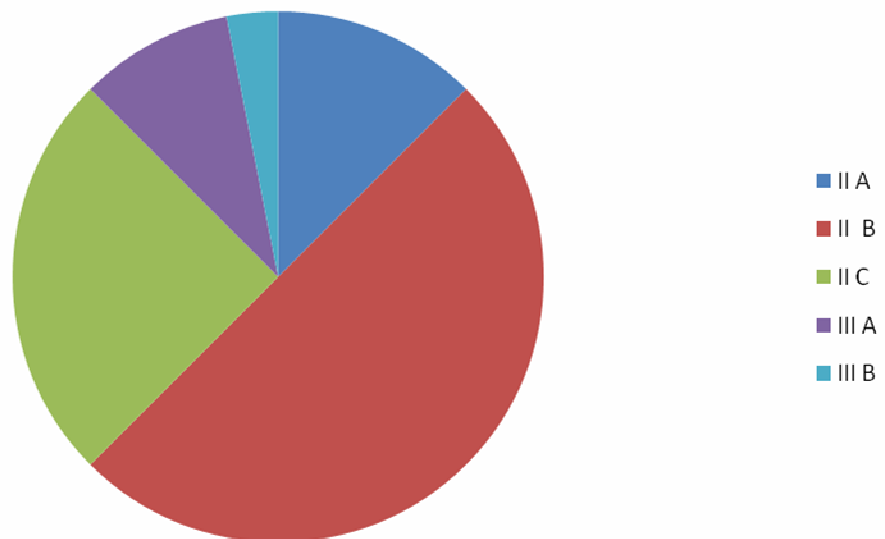




Hemostatic Methods



STAGING



CASE DETAILS

<u>S.No</u>	<u>Name & Sex (primary/recurrent)</u>	<u>Age</u>	<u>Presenting complaints</u>	<u>Clinical examination/DNE</u>	<u>Investigations (SPF-sphenopalatine foramen PPF-pterygopalatine fossa ITF- Infratemporal fossa MT-Middle turbinate)</u>	<u>Staging (Radwsky)</u>	<u>Surgical procedure(s) Hemostatic measure No.of blood transfused</u>	<u>No.of recurrences with interval from previous surgery (Yrs) (during study period)</u>	<u>Complications /Sequelae/Post op Hospital stay duration(days)</u>
1	Arun (primary)	17/ m	Nasal obstruction-4months,Epistaxis-1month.	Fleshy mass in the postnasal space	CT PNS-Enhancing vascular mass in the choanae,Right nasal cavity,SPF,PPF(Minimal)	II A	Transpalatine 3 units	Nil	Nil/(10)
2.	Srinivasan (recurrent)	13/ M	Nasal obstruction-6 months,Epistaxis 2months,Anosmia	Fleshy mass in Right nasal cavity, postnasal space	CT PNS- Enhancing mass in Rt nasal cavity,sphenoid,ethmoidSPF, PPF, ITF,extradural extension	II C	Maxillary swing approach-Twice (Ext.carotid ligation) 2 units	Once (3)	Wound gaping,Palatal fistula(Flap closure)/(20)
3	Balasundaram (Recurrent)	16/ M	Nasal obstruction-5 months,Epistaxis,Anosmia	Mass in Nasal cavity,post nasal space	CT PNS – Enhancing mass in RT.nasal cavity,Nasopharynx,RT PPF	II A	Transpalatal(3 units) Endoscopic (External carotid lig) 1 unit	Once(2)	Sinusitis,Malar anaesthesia,dacryocystitis(12,4)
4	Masthan (primary)	17/ M	Nasal obstruction, Epistaxis,Anosmia-6 months	Mass in Nasal cavity(L), choanae,	CT PNS –Enhancing mass in Lt.nasal cavity,L PPF,Choanae,Sphenoid	II B	Transnasal Endoscopic (2 units)	Nil	Synechiae,sinusitis(5)
5	Sakthy (Primary)	22/ M	Nasal obstruction-2 months,epistaxis,	Mass in Nasal cavity(R), choanae,	CT PNS – Mass in Right PPF,ITF	II C	Transnasal Endoscopic 1 unit	Nil	Nil(6)
6	Sethu (Primary)	29/ M	Nasal obstruction-4months,epistaxis,swelling of right	Mass in nasal cavity (R) ,Postnasal space,Rt	CT PNS – Mass in Rt PPF,Nasal cavity,ethmoid,Sphenoid sinus	II B	Transnasal Endoscopic 2 units	Nil	Nil(5)

CASE DETAILS

			eye	eye pushed above and out					
7	Dilliraja (Primary)	15/ M	Nasal obstruction- 1yr,epistaxis- 2 months	Mass in Lt nasal cavity attached to Middle turbinate, postnasal space	CT PNS – Mass in Right nasal cavity,Ethmoids,PPF	II B	Transnasal endoscopic, Endoscopic 2 units	Once(2)	Nil(7)
8	Jagan (Primary)	19/ M	Nasal obstruction- 5months,epi staxis	Mass in Rt. Nasal cavity & post nasal space(mist aken as AC polyp elsewhere)	CT PNS enhancing mass in the rt. nasal cavity,Nasopharynx, PPF Angiography- Rt maxillary artery is the feeding vessel and embolisation was done.	II B	Transnasal endoscopic (Embolisation) Nil transfusion	nil	Synechiae(5)
9	Bhoopath y (Recurrent)	31 /M	Nasal obstruction- 4 months,epist axis	Mass in Rt.nasal cavity, Nasophary nx	CT PNS –Mass in nasopharynx , Rt SPF,PPF	II B	Transantral- previous surgery(3 units) Transnasal endoscopic(1 Unit)	nil	Malar parassthesia(present before current surgery)(10,5)
10	Vijayakumar (primary)	17/ M	Nasal obstruction- 5 months,swelling of cheek-2 months, epistaxis- 1month	Mass in left nasal cavity(Attached to Horizontal part of MT, postnasal space Swelling over the Lt maxillary region	CT PNS- Enhancing mass in the Left nasoharynx,nasal cavity,sphenopalatine foramen,left maxilla pushed forwards and post.wall thinned out,extension into PPF ,ITF.Extradural extension to anterior cranial fossa .	III A	Maxillary swing (4 units)	Nil	Malar anaesthesia, Dacryocystitis(Endoscopic DCR done)(13)
11	Murali (recurrent)	21/ M	Nasal obstruction- 7 months, Epistaxis-2 months	Mass in the right nasal cavity attached to the horizontal	CT PNS- Enhancing mass in the right nasal cavity,sphenopalatine foramen,PPF.	II A	Endoscopic excision Nil transfusion	Nil	Nil(5)

CASE DETAILS

				portion of MT(Rt), extending to postnasal space					
12	Balaji (recurrent)	19/ M	Nasal obstruction- 4 months,epist axis-1 month,Rhino lalia	Mass in right nasal cavity attached to the septum and MT extending to the post nasal space.	CT PNS- enhancing mass in right nasal cavity, nasopharynx,SPF,PPF.	II B	Endoscopic excision (External carotid ligation) Nil transfusion	Nil	Synechiae(4)
13	Bhaskar (Primary)	19/ M	Nasal obstruction- 3 months,epist axis-3 months.	Mass in right nasal cavity attached to the MT,extending to Post nasal space.	CT PNS- Enhancing mass in right nasal cavity,Nasopharynx,SPF,PPF	II B	Endoscopic excision (Ext.carotid ligation) 1 unit	nil	Nil(5)
14	Govindara jan(primary)	13/ M	Nasal obstruction – 6 months,epist axis 2 episodes	Mass in left nasal cavity attached to MT,septum extending to postnasal space.	CT PNS- enhancing mass seen involving right nasal cavity,nasopharynx,SPF,PPF	II B	Endoscopic excision 1 unit	nil	Sinusitis(Fess done)(4)
15	Maheshw aran (primary)	20/ M	Nasal obstruction- 6 months, epistaxis-3 episodes	Mass in left nasal cavity attached to MT, septum	CT PNS- enhancing mass seen in left nasal cavity,SPF,PPF,nasopharynx	II B	Lateral rhionotomy and excision.(2 units) Endoscopic excision. Embolisation 1 unit	One(1.5)	Malar anaesthesia(12,4)

CASE DETAILS

16	Senthil (recurrent)	29/ M	Nasal obstruction- 2 months,epist axis -1 episode, headache	Mass seen in left nasal cavity arising from spenoehm oidal recess.	CT PNS- enhancing soft tissue density involving left sphenoehtmoid recess,sphenoid sinus,PPF	II B	Endoscopic excision Hypotensive anaesthesia 1 unit	nil	Nil(4)
17	Saravanan (Primary)	17/ M	Nasal obstruction- 8 months,epist axis 2 episodes, headache	Mass seen in right nasal cavity attached to MT extending to post nasal space	CT PNS- Enhancing soft tissue lesion in left nasal cavity,SPF,Nasopharynx	II A	Wilsons transpalatine approach Ext.carotid ligation 2 units	nil	Nil(12)
18	Prakash (primary)	16/ M	Nasal obstruction- 3 months,epist axis 2 episodes, rhinolalia	Mass seen in left nasal cavity attached to MT,Postnas al space	CT PNS- enhancing mass in left nasal cavity,nasopharynx,SPF,PPF	II B	Combined transantral and transpalatine approach(4 units) Endoscopic approach(1 Unit)	Nil	Palatal fistula Malar hyposthesia(22)
19	Vignesh (Primary)	17/ M	Nasal obstruction – 6 months,epist axis 4 episodes	Mass seen in right nasal cavity attached to floor,MT extending ton post nasal space	CT PNS – Enhancing mass in right nasal cavity,nasopharynx,SPF,PPF.	II B	Combined Transpalatine & endoscopic excision(Emboli sation)(2 units) Endoscopic excision (twice)(2 units,1 unit)	Two (1)	Palatal fistula (spontaneously closed) (21,4,3)
20	Vamsikrish nan (Recurrent)	16/ M	Nasal obstruction, headache-6 months,Epist axis	Mass seen in left nasal cavity, postnasal space	CT PNS- Enhancing mass in left nasal cavity,Nasopharynx,SPF,PPF,S phenoid,Extradural spread to middle cranial fossa	III A	Maxillary swing approach(Exter nal carotid ligation,3units) Endoscopic excision (2 units)	one	Malar hyposthesia, (17)
21	Saravanan (Recurrent)	19/ M	Nasal obstruction,	Mass seenin right	CT pns- Enhancing mass in Right nasal cavity,SPF,PPF,ITF.	III A	Embolisation		Death due to Massive CVA secondary to pre operative

CASE DETAILS

)		discharge-6 months,Epistaxis 4 episodes.	nasal cavity attached to MT ,post.end of septum extending to postnasal space, sphenoethmoid recess.	Posterior wall of Rt.maxilla pushed anteriorly,intracranial extradural spread to Middle cranial fossa. MRI/MRA- Enhancing hyperintense mass involving abovesaid areas with feeders from RT. ascending pharyngeal artery				embolisation.
22	Mehaboo basha (recurrent)	20/ M	Nasal obstruction, discharge Rt nasal cavity -5 months,epistaxis 2 episodes	Mass seen in Rt nasal cavity,SPF, PPF, ITF,Extradural spread to middlecranial fossa	CT pns-. Enhancing mass in Right nasal cavity,SPF,PPF,ITF. Posterior wall of Rt.maxilla pushed anteriorly,intracranial extradural spread to Middle cranial fossa.orbit MRI/MRA- Enhancing hyperintense mass involving above said areas with feeders from RT. ascending pharyngeal artery	III B	Radiotherapy		CVA with residual paresis of right upper and lower limb(managed conservatively)
23	Devanesan (recurrent)	31/ M	Nasal obstruction, discharge Rt nasal cavity -4 months.	Mass seen in Lt nasal cavity,SPF, PPF.	CT pns-. Enhancing mass in left nasal cavity,SPF,PPF. Posterior wall of Lt.maxilla pushed anteriorly. MRI/MRA- Enhancing hyperintense mass involving abovesaid areas with feeders from RT. ascending pharyngeal artery	II B	Endoscopic excision 2 units	nil	Nil(6)
24	Jaganathan (Primary)	16/ M	Nasal obstruction, discharge Rt nasal cavity -3 months,epistaxis 4 episodes	Mass seen in Rt nasal cavity,SPF, PPF	CT pns-. Enhancing mass in right nasal cavity,SPF,PPF. Posterior wall of Lt.maxilla pushed anteriorly.	II B	Endoscopic excision (Ext.carotid ligation) (1 unit)	nil	Nil(5)
25	Ranjith (primary)	17/ M	Nasal obstruction, discharge	Mass seen in Lt.nasal cavity,SPF,	CT pns-. Enhancing mass in left nasal cavity,SPF,PPF. Posterior wall of Lt.maxilla	II B	Endoscopic excision Nil transfusion	nil	Nil(7)

CASE DETAILS

			Lt.Nasal cavity,epistaxis 5 episodes	PPF	pushed anteriorly.				
26	Madhuraraj (Recurrent)	9/ M	Nasal obstruction, discharge Lt.Nasal cavity-1 month,epistaxis 5 episodes	Mass seen in the Lt.Nasal cavity attached to septum, MT extending to the sphenopalatine foramen, nasopharynx	CT PNS- Enhancing mass seen involving left nasal cavity,SPF,PPF,ITF. MRI PNS –Enhancing hyperintense lesion involving the above areas with left ascending pharyngeal artery as the main feeding vessel.	II C	Maxillary swing and excision.(EXT. carotid ligation,3 units) Endoscopic excision Nil transfusion	Recurrence (1)	Cheek anaesthesia, Dacryocystitis (underwent septal correction and endoscopic DCR) (15,5)
27	Shankar (Recurrent)	26/ M	Nasal obstruction, discharge Lt.Nasal cavity-2 months,epistaxis 5 episodes	Mass seen in the Lt.Nasal cavity attached to septum, MT extending to the nasopharynx	CT PNS- Enhancing mass seen involving left nasal cavity,SPF,PPF,. MRI PNS –Enhancing hyperintense lesion involving the above areas with left ascending pharyngeal artery as the main feeding vessel	II B	Endoscopic excision Nil transfusion	nil	Nil(4)
28	Mahesh (recurrent)	16/ M	Nasal obstruction, discharge-3 months, Headache, Epistaxis	Mass seen in the right nasal cavity attached to MT,extending to nasopharynx	CT PNS- Enhancing mass seen involving Right nasal cavity,SPF,PPF,ITF. MRI PNS –Enhancing hyperintense lesion involving the above areas with Right internal maxillary artery as the main feeding vessel	II C	Endoscopic excision (1 unit)	nil	Nil(5)
29	Natarajan (primary)	18/ M	Nasal obstruction-2 months,Nasal	Mass seen in Left nasal cavity attached	CT PNS – Enhancing lesion involving the left nasal cavity,maxillary sinus,SPF,PPF,ITF,Ethmoids.	II C	Endoscopic excision Nil transfusion	nil	Nil(3)

CASE DETAILS

[illegible]